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Journal

Pediatric Surgery International, 38(2)

ISSN

0179-0358

Authors

Kelley-Quon, Lorraine I
Shue, Eveline
Burke, Rita V
et al.

Publication Date

2022-02-01

DOI

10.1007/s00383-021-05047-1

Peer reviewed

HHS Public Access

Author manuscript

Pediatr Surg Int. Author manuscript; available in PMC 2023 February 01.

Published in final edited form as:

Pediatr Surg Int. 2022 February ; 38(2): 193–199. doi:10.1007/s00383-021-05047-1.

The Need for Early Kasai Portoenterostomy: A Western Pediatric Surgery Research Consortium Study

Lorraine I. Kelley-Quon, MD, MSHS[a,b], **Eveline Shue, MD**[a], **Rita V. Burke, PhD, MPH**[b], **Caitlin Smith, MD**[c], **Karen Kling, MD**[d], **Elaa Mahdi, MD**[a], **Shadassa Ourshalimian, MPH**[a], **Michael Fenlon, MD**[a], **Matthew Dellinger, MD**[c], **Stephen B. Shew, MD**[e], **Justin Lee, MD**[f], **Benjamin Padilla, MD**[f], **Thomas Inge, MD**[g], **Jonathan Roach, MD**[g], **Ahmed I. Marwan, MD**[g], **Katie W. Russell, MD**[h], **Romeo Ignacio, MD**[d], **Elizabeth Fialkowski, MD**[i], **Amar Nijagal, MD**[j], **Cecilia Im, BS**[j], **Kenneth S. Azarow, MD**[i], **Daniel J. Ostlie, MD**[f], **Kasper Wang, MD**[a]

[a]Division of Pediatric Surgery, Children's Hospital Los Angeles and the Keck School of Medicine of the University of Southern California, Los Angeles, CA;

[b]Department of Preventive Medicine, Keck School of Medicine, University of Southern California, Los Angeles, CA;

[c]Division of Pediatric General and Thoracic Surgery, Seattle Children's Hospital, Seattle WA and Department of Surgery, University of Washington School of Medicine, Seattle, WA;

[d]Department of Pediatric Surgery, Rady Children's Hospital San Diego, University of California School of Medicine, San Diego, CA;

[e]Division of Pediatric Surgery, Department of Surgery, Stanford University School of Medicine, Palo Alto, CA;

[f]Division of Surgery, Phoenix Children's Hospital, Phoenix, AZ;

[g]Department of Surgery, University of Colorado Denver School of Medicine and Children's Hospital of Colorado, Aurora, CO;

[h]Division of Pediatric Surgery, University of Utah and Primary Children's Hospital, Salt Lake City, UT;

[i]Division of Pediatric Surgery, Department of Surgery, Oregon Health & Science University, Portland, OR;

Corresponding Author: Lorraine I. Kelley-Quon, MD, MSHS, FACS, FAAP, Children's Hospital of Los Angeles, Division of Pediatric Surgery, 4650 Sunset Blvd. MS #100, Los Angeles CA 90027, Phone: 323.397.8539 Fax: 323.361.3534 lkquon@chla.usc.edu.

Author Contributions: All authors contributed to the study conception and design. Material preparation, data collection, and analysis were performed by Lorraine Kelley-Quon, Rita Burke, Elaa Mahdi, and Shadassa Ourshalimian. The first draft of the manuscript was written by Lorraine Kelley-Quon and all authors commented on previous versions of the manuscript. All authors read and approved the final manuscript.

Consent to Participate: Institutional Review Board (IRB) approval was obtained independently by each member institution. All member institution IRBs approved a waiver of consent to collect medical record data.

[j]Division of Pediatric Surgery, University of California, San Francisco, San Francisco, CA

Abstract

Purpose: The purpose of this study was to investigate factors impacting transplant-free survival among infants with biliary atresia.

Methods: A multi-institutional, retrospective cohort study was performed at nine tertiary-level children's hospitals in the United States. Infants who underwent Kasai portoenterostomy (KP) from January 2009-May 2017 were identified. Clinical characteristics included age at time of KP, steroid use, surgical approach, liver pathology, and surgeon experience. Likelihood of transplant-free survival (TFS) was evaluated using logistic regression, adjusting for patient and surgeon-level factors. Secondary outcomes at 1-year included readmission, cholangitis, reoperation, mortality, and biliary clearance.

Results: Overall, 223 infants underwent KP, and 91 (40.8%) survived with their native liver. Mean age at surgery was 63.9d (±24.7d). At 1-year, 78.5% experienced readmission, 56.9% developed cholangitis, 3.8% had a surgical revision, and 5 died. Biliary clearance at 3 months was achieved in 76.6%. Controlling for patient and surgeon-level factors, each additional day of age toward operation was associated with a 2% decrease in likelihood of TFS (OR 0.98, 95% CI 0.97–0.99).

Conclusion: Earlier surgical intervention by Kasai portoenterostomy at tertiary-level centers significantly increases likelihood for TFS. Policy-level interventions to facilitate early screening and surgical referral for infants with biliary atresia are warranted to improve outcomes.

Keywords

transplant-free survival; liver transplant; biliary atresia; Kasai portoenterostomy

Introduction

Biliary atresia (BA) is a congenital fibro-obliterative cholangiopathy of intra- and extrahepatic bile ducts that involves progressive liver fibrosis which results in cirrhosis and liver failure if left untreated [1,2]. Although rare, presenting with an incidence of 4 per 100,000 live births in the United States, it is the most common cause of end-stage liver disease in children and continues to be the leading indication for pediatric liver transplantation [3]. The Kasai portoenterostomy (KP) is a highly technical surgery performed to try to restore bile flow in the liver of patients with BA [4]. However, only 30–50% of infants attain successful, long-term biliary drainage after a portoenterostomy [1,3,5]. A significant challenge is early detection and diagnosis of biliary atresia. Evidence supports that early surgical treatment by 30-days of life has the greatest likelihood of slowing or preventing the need for liver transplant [6,7]. However, in the United States because biliary atresia is challenging to detect early, surgical intervention often occurs later [8].

A successful KP has been associated with improved native-liver survival and delayed requirement for transplant [9]. Patient prognostic factors associated with KP success include liver histology, biliary remnant anatomy, age at time of operation, presence of

ascites, and episodes of cholangitis [1,5,10–13]. Several European studies have reported that centralization of BA care and increased institutional volume are associated with higher jaundice clearance rates and native-liver survival [14–16]. However, an analysis of outcomes after KP for Canadian infants failed to find a significant association between institutional caseload and likelihood of liver transplant [17]. Similar studies have not been performed in the United States.

For several complex adult operations, higher individual surgeon case volumes are associated with improved outcomes [18–20]. Whether individual surgeon volume versus patient-level factors impact outcomes after KP is unknown. Therefore, we investigated pertinent clinical factors in addition to individual surgeon experience of KP to determine the relative impact on transplant-free survival among infants with biliary atresia.

Methods

Study Design and Data Collection

This study is a retrospective cohort study conducted by the Western Pediatric Surgical Research Consortium (WPSRC) member hospitals. WPSRC is a multi-institutional pediatric surgical collaborative of nine tertiary-level children's hospitals across the Western United States. This consortium is committed to advancing the care of infants and children through evidence-based research. Participating WPSRC hospitals queried medical records for all infants who underwent KP from January 1, 2009 to May 19, 2017 for the diagnosis of biliary atresia. Clinical and patient demographics were extracted, de-identified, and managed through the HIPAA compliant Web application, Research Electronic Data Capture (REDCap). Institutional Review Board approval was obtained independently by each member institution.

Patient Characteristics and Outcomes

Patient characteristics of interest included sociodemographic variables (sex, race, ethnicity, and insurance status). Clinical characteristics of interest included age at time of KP, type of biliary atresia, other preoperative congenital anomalies, and transplant-free survival (TFS). Primary postoperative outcome evaluated was likelihood of TFS, defined as a patient who was not listed for an orthotopic liver transplant, did not receive an orthotopic liver transplant, and/or who survived after KP at 1-year after operation. Secondary outcomes at 1-year included overall incidence of readmissions, cholangitis, surgical revision, mortality, and attaining biliary clearance (defined as a total bilirubin < 2.0 mg/dl at 3 months postoperatively). The cutoff for biliary clearance was selected as previous studies have highlighted 3-month biliary clearance as significantly associated with transplant-free survival [21]. If an infant transferred care from one institution within the consortium to another institution within the consortium, their outcomes were tracked longitudinally. Infants within each hospital's region with biliary atresia who underwent KP at a hospital outside of the consortium were not captured in this analysis.

Definition of Surgeon Volume and Surgeon Experience

Surgeon volume was defined as the total number of *prior* KPs performed by the surgeon for an index operation during the study period. A surgeon entering the study would be considered to have a cumulative volume of zero when performing the first KP, on the second KP his or her cumulative volume would equal one, and so on. If a physician assisted as secondary or tertiary surgeon, we included that operation in their cumulative surgeon volume. Individual surgeon volume was assessed as the study progressed and cumulative surgeon volume increased for each KP performed.

Statistical Analysis

Categorical variables were reported as frequencies and percentages and continuous variables were described by mean and standard deviation. Patient demographics were compared using bivariate analyses. Categorical and continuous variables were assessed by using χ^2 or Fisher exact tests and Two-Sample t-test, respectively. Continuous variable distributions were assessed for normality by using histograms and Q-Q plots. Predicted probabilities of TFS by age at time of KP were examined using penalized B-spline fit line of increasing age at time of KP. A multivariable logistic regression model was used to evaluate cumulative surgeon volume and overall likelihood of TFS. Infants who were lost to follow-up (n=16) were excluded from our primary analysis. The regression models adjusted for age in days at time of KP, steroid use (any), documented hilar dissection (dissection above the portal plate into the substance of the liver, a non-standard approach), and fibrosis present on pathology. Comparison of rates of TFS between individual centers was also evaluated.

Missing data were handled by multiple imputation using chained equations to construct 40 imputed datasets with the assumption that categorical variables for postoperative steroid use, hilar dissection, and fibrosis present on pathology were missing at random. Auxiliary variables used to inform the full conditional imputations included patient sex, race, ethnicity, insurance status, type of biliary atresia, transplant-free survival status, age at KP, and study site. All analyses were conducted with two-sided significance, $\alpha = 0.05$. Data were analyzed by using SAS software 9.4 (SAS Institute, Inc, Cary, North Carolina) and StataCorp (StataCorp LLC, College Station, Texas).

Results

The final cohort, excluding infants who were lost to follow-up, was 223 infants with biliary atresia who underwent KP at the nine WPSRC hospitals over an 8-year period (Supplemental Figure 1). Infants who underwent KP were predominantly female (63.2%), white (62.8%), with little difference between private and public insurance (48.9 vs 50.7%) (Table 1). Overall, mean age at time of KP was 63.9 0 (±24.7) days. Fibrosis was present on liver pathology among 86.6% of infants. Atresia of hepatic duct (Type III) was the most common pathological anatomy present (73.1%). Other congenital anomalies were present among 38 (17.0%) of infants, with the most common anomalies being cardiac (n=18, 47.4%). KP caseloads of 68 surgeons were evaluated, with cumulative individual surgeon volumes ranging from 1–26 at the conclusion of the study period (see Supplemental Figure 2). Surgeons applied a hilar dissection above the portal plate in 32.7% of cases and

used postoperative steroids in 29.6% of cases. The lowest volume center in the consortium performed 8 KP during the study and the highest volume center performed 52. There were no significant differences between rates of TFS between individual hospitals within the consortium.

Over the course of the study period, 120 infants (53.8%) underwent orthotopic liver transplant (OLT) and 12 died (5.4%) leaving 91 (40.8%) infants surviving with their native liver after KP. Of those who underwent OLT, the mean number of days from KP to OLT listing was 289.4 (± 492.1) days and from KP to OLT was 428.9 (±527.5) days. Overall, 116 (96.7%) infants received an OLT within five years of KP and all deaths for patients not listed or transplanted occurred within 5 years of KP. The indication for transplantation was liver failure due to fibrosis in all infants. Few statistical differences were detected among infants who survived with their native liver versus those who underwent OLT, except for age at time of KP. Infants who survived with their native liver were considerably younger at the time of their KP (57.0 vs. 68.6 days, $p<0.001$) (Table 1). Predicted probabilities of TFS by age at time of KP showed a near-linear relationship with age at time of surgery (Figure 1).

On logistic regression analysis, surgeon cumulative case volume was not significantly associated with improved TFS (Table 2). Similarly, TFS was not improved with postoperative steroid use, surgical approach, or when noting fibrosis on pathology at time of KP. However, age in days at time of KP (OR 0.98, 95% CI 0.97–0.99) demonstrated a significant impact on TFS while holding other factors constant. Ultimately, each one day increase in age at time of surgery was associated with a 2% decrease in likelihood of TFS. There was no difference in TFS outcome between surgeons with the largest surgical case volume (>10 cases) and surgeons with lowest (<4 cases) case volume, ($p = 0.919$).

On evaluation of secondary outcomes for our overall cohort; 78.5% infants experienced one-year readmission, 56.9% had symptoms of cholangitis, 3.8% had a KP revision (Table 3). Of the overall cohort, 77 children who had not undergone OLT in the 3 months following KP had a total bilirubin reported at 3 months. A total bilirubin <2.0, indicating early biliary clearance, occurred in 59 (76.6%) infants at 3 months. All-cause mortality was 5.0%. There was no difference between centers for one year readmission ($p=0.323$) or biliary clearance ($p=0.235$) but there were differences in the prevalence of cholangitis ($p<0.001$).

Discussion

To our knowledge, the present study is the largest, multi-institutional retrospective U.S. cohort study to evaluate the relationship between patient and surgeon-level factors and outcomes after KP. The study is robust in its analysis as we were able to follow individual surgeons at multiple children's hospitals over time and allow for an individual surgeon to progress in cumulative case volume over time. Our findings indicate that the strongest predictor associated with overall likelihood of TFS is age at time of KP. This study provides new evidence in support of early screening for biliary atresia, referral to a tertiary-level children's hospital, and prompt operative intervention.

Considerable variability in age at KP for biliary atresia has been noted in United States children's hospitals [22]. In the present study, age at time of KP was the strongest predictor of TFS. On regression analysis, each one day increase in age was associated with a 2% decrease in the likelihood of TFS (OR 0.98, 95% CI 0.97–0.99). Similar findings have been previously reported in other international cohorts [5,12] and underscore the rapidly progressive pathophysiology inherent in biliary atresia. Of note, the mean age at time of KP in the present study was 64 days, while other groups reporting on successful outcomes after centralization of surgical care report mean ages of 53 days [14,15]. This finding underscores the challenge faced by clinicians in the United States to identify biliary atresia early, as a 1-month well-child visit (which would be an optimal time for screening and/or identification of persistent jaundice) is not typically offered to otherwise healthy infants [4].

Early referral and intervention may also be improved with standardized institution of newborn screening for biliary atresia in the United States. Recent studies in the United States have demonstrated that early screening for biliary atresia can be achieved using direct or conjugated bilirubin measurements soon after birth. Utilizing direct or conjugated screening had a sensitivity of 100%, a specificity of 99.9%, a positive predictive value of 5.9%, and a negative predictive value of 100.0%. Furthermore, in a pre-post study, the mean age infants underwent the KP was significantly younger following the implementation of screening (56 days vs 36 days, P=0.004) [6].

Previous studies in Europe highlight improved outcomes and lower rates of liver transplantation after KP with regionalization of surgical care to specialty centers. However, when looking at annual center volumes of European regional centers, overall case volume did not increase substantially after regionalization. In McKiernan et al.'s review of outcomes after regionalization of care in the United Kingdom and Ireland, high-volume centers were defined as hospitals performing more than 5 portoenterostomies per year [14]. In Finland, individual surgeon median annual caseload increased from 1 to 3.5 after centralization [16]. However, both the U.K. and Finland witnessed a substantial drop in overall transplant rates associated with regionalization of care for infants with biliary atresia. These results imply that the change in referral patterns and the multidisciplinary surgical care and follow-up received at these specialized centers may improve outcomes after KP. The landscape of health care delivery in the U.S. varies widely from health care delivered in European countries. Outcomes for infants with biliary atresia very well may improve in the U.S. with more expedient referral, centralization of care, and early detection.

The present study is limited by the retrospective nature of review. Outcomes for infants that received postoperative care at hospitals outside of the WPSRC were unable to be adequately followed, although this represented only 7% of the overall cohort. In addition, we were unable to account for the case volume of an individual surgeon prior to 2009 and thus unable to address the impact of a 20 to 30-year surgical career on outcomes. However, we did adjust for the total number of years a surgeon was in practice prior to the start of the cohort in 2009 as a surrogate for surgeon experience. The findings of this present study are strengthened by the eight-year period of investigation allowing for an individual surgeon to progress in cumulative case volume and experience. The findings and outcomes conveyed are also limited by the use of data from only tertiary-level children's hospitals, and therefore

cannot be extrapolated to KPs that may be performed in less specialized hospitals globally. Ultimately, these data underscore the importance of early referral, diagnosis, and surgical treatment of biliary atresia.

Conclusion

Age at time of Kasai portoenterostomy appears to be most strongly associated with likelihood of survival with native liver for infants with biliary atresia. These findings highlight the need for early diagnosis of biliary atresia and expedient referral for surgical management. Policy-level interventions to facilitate early screening and surgical referral for infants with biliary atresia are warranted to improve outcomes.

Supplementary Material

Refer to Web version on PubMed Central for supplementary material.

Funding/Support:

Dr. Kelley-Quon is supported by grant KL2TR001854 from the National Center for Advancing Translational Science (NCATS) of the U.S. National Institutes of Health. The content is solely the responsibility of the authors and does not necessarily represent the official views of the National Institutes of Health. The remaining authors have no relevant financial or non-financial interests to disclose.

Conflicts of Interest and Source of Funding:

The authors have no financial relationships relevant to this article or potential competing interests to disclose.

Abbreviations:

BA	biliary atresia
HIPAA	Health Insurance Portability and Accountability Act
KP	Kasai portoenterostomy
OLT	orthotopic liver transplant
REDCap	Research Electronic Data Capture
TFS	transplant free survival
WPSRC	Western Pediatric Surgical Research Consortium

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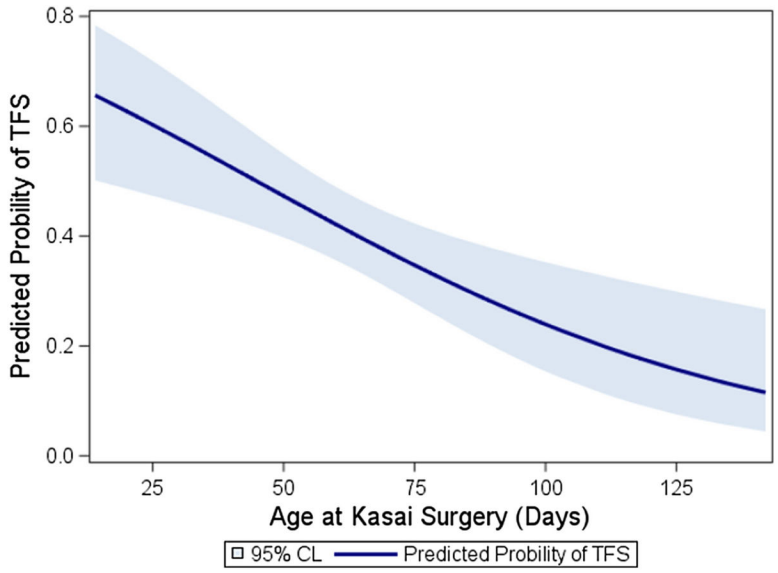

Figure 1.
Predicted probabilities of Transplant-free Survival (TFS) by Age at Time of Kasai Portoenterostomy.

Table 1.

Cohort demographics and clinical characteristics by transplant-free survival

	Overall N=223 (%)	Transplant-Free Survival N=91 (%)	Listed for transplant, transplanted, or expired N = 132 (%)	P-value
Female Sex	141 (63.2)	54 (59.3)	87 (65.9)	0.317
Race				0.416
American Indian	1 (0.5)	0 (0.0)	1 (0.8)	
Asian	14 (6.3)	6 (6.6)	8 (6.1)	
Black	7 (3.1)	3 (3.3)	4 (3.0)	
Multiple	21 (9.4)	12 (13.2)	9 (6.8)	
Unknown	40 (17.9)	12 (13.2)	28 (21.2)	
White	140 (62.8)	58 (63.7)	82 (62.1)	
Ethnicity				0.629
Hispanic	62 (27.8)	25 (27.5)	37 (28.0)	
Non-Hispanic	150 (67.3)	63 (69.2)	87 (65.9)	
Unknown	11 (4.9)	3 (3.3)	8 (6.1)	
Insurance				0.579
Other	1 (0.4)	0 (0.0)	1 (0.8)	
Private	113 (50.7)	44 (48.3)	69 (52.3)	
State	109 (48.9)	47 (51.7)	62 (47.0)	
Age in days at surgery, mean (±SD)	63.9 (±24.7)	57.0 (±23.1)	68.6 (±24.7)	<0.001
Fibrosis Present on Pathology[a]	193 (86.6)	80 (87.9)	113 (85.6)	0.631
Type of Biliary Atresia				0.225
Type I	43 (19.3)	12 (13.2)	31 (23.5)	
Type II	15 (6.7)	7 (7.7)	8 (6.1)	
Type III	163 (73.1)	71 (78.0)	92 (69.7)	
Cystic Biliary Atresia	2 (0.9)	1 (1.1)	1 (0.8)	
Congenital Anomalies	38 (17.0)	17 (18.7)	21 (15.9)	0.588
Cardiac	18 (47.4)	7 (41.2)	11 (52.4)	
Asplenia/polysplenia	5 (13.2)	2 (11.8)	3 (14.3)	
Sacral agenesis	1 (2.6)	0 (0.0)	1 (4.8)	
Heterotaxy	4 (10.5)	2 (11.8)	2 (9.5)	
Situs inversus	2 (5.3)	0 (0.0)	2 (9.5)	
Cleft lip/palate	2 (5.3)	2 (11.8)	0 (0.0)	
Renal pelviectasis	3 (7.9)	2 (11.8)	1 (4.8)	
Other	21 (55.3)	12 (70.6)	9 (42.9)	
Hilar Dissection[b]	73 (32.7)	31 (34.1)	42 (31.8)	0.760
Postoperative Steroids[c]	66 (29.6)	27 (29.7)	39 (29.6)	0.989
Number of clinic visits 6 months after surgery, mean (±SD)	1.6 (±1.6)	1.7 (±1.3)	1.6 (±1.8)	0.321

[a] n=214

[b] n=187

[c] n=205

Table 2.

Multivariable Logistic Regression of Likelihood of Transplant-free Survival

Parameter	OR	95% CL		p-value
Age in days at Kasai portoenterostomy	0.98	0.97	0.99	<0.001
Postoperative steroid use	1.00	0.53	1.87	0.998
Hilar dissection	0.65	0.37	1.17	0.151
Fibrosis on pathology	1.38	0.53	3.57	0.505
Surgeon cumulative case volume	0.97	0.92	1.02	0.262

Table 3.

Secondary Outcomes

	N (%)
One-year readmission	175 (79.2)
Biliary Clearance[a]	59 (76.6)
Cholangitis	129 (57.9)
Kasai revision[b]	7 (3.1)
Death, all causes[b]	12 (5.4)

[a] defined as total bilirubin < 2.0 mg/dl at 3 months postoperatively for non-OLT patients (N=77)