

Clinical outcomes after Kasai portoenterostomy in biliary atresia: role of early surgery and liver surface telangiectasia in a resource-limited setting

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ABSTRACT

Aims: Biliary atresia (BA) remains a leading cause of pediatric liver failure worldwide, with outcomes heavily influenced by the timing of intervention and healthcare resources. In low- and middle-income countries, delayed diagnosis and limited access to specialized care contribute to poorer outcomes following Kasai portoenterostomy (KPE). This study aimed to evaluate clinical outcomes and identify prognostic factors in infants undergoing KPE at a tertiary pediatric center.

Methods: This retrospective observational study included all consecutive infants with BA who underwent KPE at our center between January 2022 and December 2025. Data on demographic characteristics, preoperative biochemical parameters, intraoperative findings, and postoperative outcomes were collected. Survival and jaundice clearance were assessed at 3, 6, and 12 months. Associations between clinical variables and outcomes were analyzed using the Mann-Whitney U test, the Chi-square test, or Fisher's exact test, with $p < 0.05$ considered statistically significant.

Results: A total of 52 infants were included, with a median age at operation of 71 days. Liver surface telangiectasia was present in 63.5% of patients. The 30-day mortality was 7.7%, and survival rates at 3 months, 6 months, and 1 year were 65.4%, 51.9%, and 48.1%, respectively. Jaundice clearance at 6 months was achieved in 34.6% of patients. Younger age at operation was significantly associated with jaundice clearance (median 62 vs 72 days, $p = 0.046$). Preoperative direct bilirubin was significantly higher among non-survivors ($p = 0.028$). Liver surface telangiectasia showed a trend toward poorer jaundice clearance ($p = 0.055$). Kaplan-Meier analysis demonstrated a progressive decline in survival, particularly within the early postoperative period.

Conclusion: Outcomes of BA following KPE remain suboptimal in this setting, largely due to delayed presentation and advanced disease at surgery. Early surgical intervention is the most important modifiable factor associated with improved bile drainage, while elevated direct bilirubin and liver surface telangiectasia may indicate poorer prognosis. Strengthening early diagnosis and referral pathways is essential to improving outcomes in resource-limited settings.

Keywords: Biliary atresia, Kasai portoenterostomy, jaundice clearance, survival analysis, liver fibrosis, pediatric surgery

INTRODUCTION

Biliary atresia (BA) is a progressive fibro-obliterative cholangiopathy of infancy characterized by inflammation and obliteration of the extrahepatic biliary tree, leading to persistent cholestasis, biliary cirrhosis, and ultimately liver failure if untreated. It represents the most common indication for pediatric liver transplantation worldwide and continues to pose significant clinical and research challenges despite advances in diagnosis and management.¹

In low- and middle-income countries (LMICs), the management of BA faces several unique challenges, including delayed diagnosis, limited awareness among primary healthcare

providers, restricted access to specialized hepatobiliary surgical services, and limited availability of pediatric liver transplantation. These factors often result in late presentation and reduced success rates following Kasai portoenterostomy (KPE). Studies from several LMICs have demonstrated that many infants undergo surgery beyond the optimal age for intervention, which significantly affects postoperative bile drainage and long-term native liver survival.²

While Kasai Porto enterostomy is the only hope for these patients in LMICs, informed decision-making is sometimes challenging in the pre- and post-operative period. Aside from



advanced age, biochemical markers, and histology, there is no universally accepted gross liver finding that can predict poorer outcome or advanced liver damage in these patients. Zhang et al.³ reported that the hepatic subcapsular telangiectasia sign was present in all infants with BA and absent in non-BA cholestatic disorders, with 100% sensitivity and specificity for diagnosis; however, they also noted that this could be seen even in very young infants without obvious cirrhosis, indicating that the sign is not simply a marker of advanced fibrosis.

Despite these challenges, evidence suggests that meaningful clinical benefit can still be achieved when KPE is performed in specialized centers with appropriate surgical expertise and postoperative care. Evaluating institutional outcomes in LMIC settings is therefore essential for identifying gaps in care, improving referral pathways, and optimizing management strategies for children with BA.^{2,4}

The objective of this study was to evaluate the clinical outcomes of infants with BA undergoing KPE at a tertiary pediatric center. In addition, the study aims to analyze postoperative outcomes and identify age at surgery and liver surface telangiectasia as prognostic factors following KPE.

METHODS

The study was conducted in accordance with the ethical principles set forth in the Declaration of Helsinki. Ethical approval was obtained from the Ethics Review Board of the Bangladesh Shishu Hospital and Institute (Date: 15.01.2026, Decision No: Admin/BSHI/2026/207). Patient confidentiality was maintained throughout the study, and all data were anonymized prior to analysis.

This retrospective observational study was conducted at a tertiary pediatric referral center providing specialized pediatric surgical services. The study evaluated the clinical outcomes of infants diagnosed with BA who underwent KPE at this institution.

All consecutive infants diagnosed with BA who underwent KPE by the same surgery team between January 2022 and December 2025 were included in the study. Patients with incomplete clinical data and those who did not undergo KPE (including patients who were not fit for surgery and parents who refused surgery) were excluded.

Data were collected retrospectively from hospital medical records, operative notes, and follow-up documentation using a structured data collection form. The following variables were recorded: age at surgery, preoperative biochemical parameters, intraoperative findings (including liver surface telangiectasia), postoperative biochemical parameters, jaundice clearance, and survival outcomes. Liver surface telangiectasia was defined as the macroscopic presence of subcapsular vessels on the liver surface.

Statistical Analysis

Primary outcomes were survival with native liver and jaundice clearance at 3, 6, and 12 months. Serum total bilirubin below 2.0 mg/dl at 6 months after surgery was considered jaundice clearance. Continuous variables were expressed as mean or

median (range), and categorical variables as frequencies and percentages. Comparisons were performed using the Mann–Whitney U test, Chi-square test, or Fisher's exact test as appropriate. A p-value <0.05 was considered statistically significant.

RESULTS

A total of 52 infants with BA who underwent KPE during the study period were included in the analysis. **Table 1** summarizes the independent variables. The mean age at operation was 78.3 days, with a median of 71 days (range 33–155 days). Liver surface telangiectasia (**Figure 1**) was present in 33 patients (63.5%).

Table 1. Comparison of variables between survivors and non-survivors

Variable	Survivors (n=25)	Non-survivors (n=27)	p-value
Male sex, n (%)	14 (56.0%)	18 (66.7%)	0.614
Age at operation (days), median (range)	68 (33–155)	72 (33–155)	0.077
Preoperative total bilirubin (mg/dl), mean	12.2	12.9	0.667
Preoperative direct bilirubin (mg/dl), mean	7.86	9.69	0.028
Liver surface telangiectasia present, n (%)	10 (40.0%)	19 (70.4%)	0.027

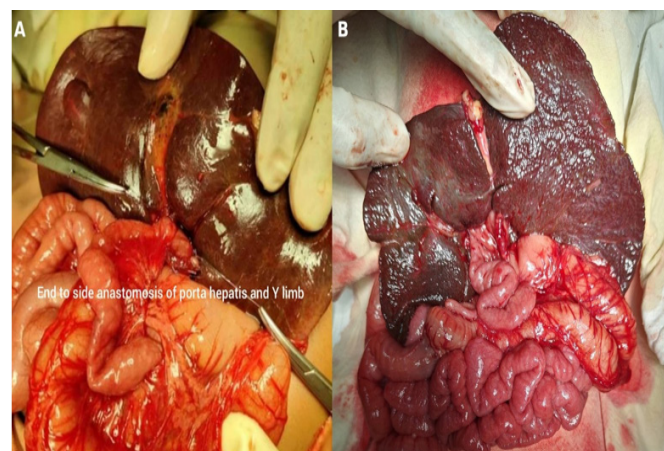


Figure 1. A- Liver without surface telangiectasia; B- Liver with surface telangiectasia

The 30-day mortality was 7.7% (4/52). These patients were categorized as non-clearance of jaundice. The mean preoperative total bilirubin was 12.6 mg/dl, decreasing to 7.8 mg/dl at 2 weeks after surgery among 48 patients (excluding 4 patients with 30-day mortality). Mean direct bilirubin decreased from 8.8 mg/dl preoperatively to 5.4 mg/dl at two weeks. At three months, mean total bilirubin decreased further to 5.1 mg/dl, with a mean direct bilirubin of 4.0 mg/dl, demonstrating progressive biochemical improvement among surviving patients. **Table 2** summarizes the survival outcomes and jaundice clearance.

Those who achieved jaundice clearance underwent surgery at a significantly younger age than those who did not. The median age at operation was 62 days in the jaundice-clearance group, compared with 72 days in the non-clearance group

**Table 2.** Survival outcomes and jaundice clearance

Outcome	Value
30-day mortality	4/52 (7.7%)
3-month survival	34/52 (65.4%)
6-month survival	27/52 (51.9%)
1-year survival	25/52 (48.1%)
Jaundice clearance at 6 months	18/52 (34.6%)

($p=0.046$). There was no significant difference in preoperative total bilirubin (12.34 vs 12.50 mg/dl, $p=0.942$) or preoperative direct bilirubin (8.06 vs 8.14 mg/dl, $p=0.866$) between the two groups.

Liver surface telangiectasia showed a borderline association with jaundice clearance. Jaundice clearance occurred in 11 of 13 patients (84.6%) without telangiectasia compared with 7 of 15 patients (46.7%) with telangiectasia, $p=0.055$ (Table 3).

Table 3. Factors associated with jaundice clearance

Variable	Cleared (mean/median)	Not cleared (mean/median)	p-value
Age at operation (days)	62.5/62	88.6/72	0.046
Total bilirubin (mg/dl)	12.34/12.5	12.50/13	0.942
Direct bilirubin (mg/dl)	8.06/8.5	8.14/7.5	0.866
Telangiectasia (yes vs no)	7/15 vs 11/13	-	0.055

Multivariable Logistic Regression for Jaundice Clearance

A multivariable logistic regression model was performed, including age at operation, preoperative direct bilirubin, and liver surface telangiectasia. None of the variables remained independently associated with jaundice clearance in the adjusted model (Table 4).

Table 4. Regression analysis

Variable	OR	95% CI	p-value
Age at operation (per day)	1.015	0.954–1.081	0.631
Preoperative direct bilirubin (mg/dl)	0.585	0.262–1.308	0.191
Liver surface telangiectasia	0.23	0.03–2.05	1.000

OR: Odds ratio, CI: Confidence interval

Kaplan–Meier analysis demonstrating overall survival of children with BA following KPE over a 12-month follow-up period. Survival probability declined progressively during the first year, with the greatest reduction occurring in the early postoperative months (Figure 2).

DISCUSSION

This study evaluates the clinical outcomes and prognostic factors following KPE in a cohort of infants with BA from a tertiary pediatric center in Bangladesh. The findings highlight the impact of early surgical intervention, biochemical parameters, and intraoperative findings on postoperative outcomes in a resource-limited setting.

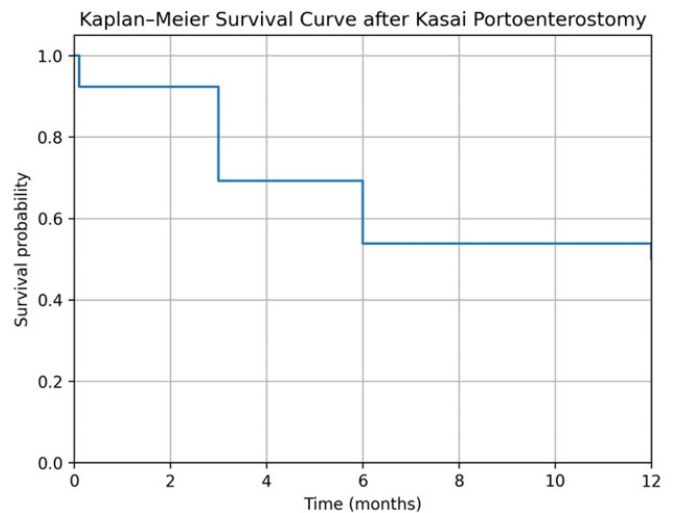


Figure 2. Kaplan–Meier survival analysis demonstrating overall survival of children with biliary atresia following Kasai portoenterostomy over a 12-month follow-up period

The overall one-year survival with native liver in this cohort was 48.1%, which is lower than outcomes reported from high-income countries, where survival rates often exceed 60–70% following centralized care and early intervention.^{4,9} However, the outcomes observed in this study are comparable to reports from other LMICs, where delayed presentation and limited access to specialized care remain significant challenges.^{2,5,10,11} This reinforces the persistent disparity in outcomes for BA between high-resource and low-resource settings.

Age at surgery emerged as an important determinant of postoperative outcome, particularly in relation to jaundice clearance. Patients who achieved jaundice clearance underwent surgery at a significantly younger age compared to those who did not. This finding is consistent with multiple studies demonstrating that early KPE—ideally before 60 days of life—is associated with improved bile drainage and long-term native liver survival.^{4,9} Although age did not show a statistically significant association with survival in this cohort, the observed trend likely reflects the limited sample size and reduced statistical power.

Preoperative direct bilirubin was significantly higher among non-survivors, suggesting that greater cholestatic burden at presentation may reflect more advanced disease and poorer hepatic reserve. Similar associations have been reported in previous studies, where elevated bilirubin levels correlate with worse postoperative outcomes and increased risk of liver failure.^{6,8} However, total bilirubin did not show a significant association, indicating that direct bilirubin may be a more sensitive marker of disease severity in this context.

A key and innovative aspect of this study is assessing liver surface telangiectasia as a potential prognostic marker. Although initially identified as a diagnostic indicator of BA with high sensitivity and specificity,³ its prognostic value remains uncertain. Although liver surface telangiectasia was associated with poorer outcomes on univariable analysis, its effect could not be reliably estimated in adjusted modeling due to sparse data and model instability. This likely reflects



the small sample size and quasi-separation, rather than the absence of a true association. The observed trend is clinically important and warrants further study.

The rate of jaundice clearance at six months in this cohort was 34.6%, which is lower than rates reported in high-income settings but comparable to those reported in other LMIC studies.¹⁰⁻¹³ Jaundice clearance is a key early surrogate marker of successful biliary drainage and has been consistently associated with improved long-term outcomes, including native liver survival.^{7,12} The relatively low clearance rate observed in this study may reflect delayed presentation, advanced disease at the time of surgery, and limitations in postoperative care.

The Kaplan–Meier survival analysis demonstrated a progressive decline in survival during the first year, with the most significant drop occurring in the early postoperative period. This pattern is consistent with previous reports, where early postoperative mortality is often related to sepsis, cholangitis, or liver failure.^{2,6} The absence of loss to follow-up in this study strengthens the validity of the survival analysis and enhances the reliability of outcome assessment.

Overall, the findings of this study emphasize the critical importance of early diagnosis, timely referral, and surgical intervention in improving outcomes for BA in LMIC settings. In addition, the potential role of intraoperative findings, such as liver surface telangiectasia, as prognostic indicators warrant further research.

Limitations

The absence of histopathological correlation of the liver surface telangiectasia restricted deeper evaluation of disease severity, and long-term outcomes beyond one year could not be assessed. Additionally, as a tertiary referral center, we may be subject to referral bias, with a higher proportion of advanced cases included.

Recommendations

Despite these limitations, the study provides valuable insights into outcomes in BA in a low-resource setting. Future efforts should focus on establishing national screening and early referral programs to facilitate timely diagnosis and intervention. Increasing awareness among primary healthcare providers, developing centralized management pathways, and promoting multicenter collaborative studies are essential to improve outcomes. Further research is also warranted to explore the prognostic significance of intraoperative findings such as liver surface telangiectasia and to optimize postoperative care strategies.

CONCLUSION

BA continues to be associated with suboptimal outcomes in low-resource settings, primarily due to delayed presentation and advanced disease at the time of surgery. Early age at KPE remains the most important modifiable factor associated with improved bile drainage, while elevated preoperative direct bilirubin and the presence of liver surface telangiectasia may indicate more advanced disease and poorer prognosis.

Strengthening early detection strategies and referral pathways is essential to improve clinical outcomes in this population.

ETHICAL DECLARATIONS

Ethics Committee Approval

Ethical approval was obtained from the Ethics Review Board of the Bangladesh Shishu Hospital and Institute (Date: 15.01.2026, Decision No: Admin/BSHI/2026/207).

Informed Consent

As this was a retrospective study, formal written informed consent was not required and was therefore not obtained.

Peer Review Process

This manuscript was subject to external peer review.

Conflict of Interest

The authors declare no conflicts of interest related to this study.

Financial Disclosure

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Author Contributions

Concept: MSH; Design: MSH; Control: AR; Resources: MSH; UFKBB, RM, Materials: MSH, UFKB; Data Collection and/or Processing: UFKBB, RM; MSH; Analysis and/or Interpretation: MSH, SMNI; Literature Review: MSH, RM; Writing the Article: MSH; Critical Review: MSH; UFKBB, RM, SMNI, AR.

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