Published by
Saidu Medical College

OPEN ACCESS

ISSN (P): 1819-4583
ISSN (E): 3007-9470

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Citation:
Hussain F, Jan K, Ahmed A, Rahim S, Ahmad S, Haq IU. Short-Term Clinical Outcomes of Kasai Porto-Enterostomy for Biliary Atresia: A Seven-Year Single-Center Experience from a Tertiary Care Hospital in Pakistan. *J Saidu Med Coll.* 2026;16(3):483–490.

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Funding:
The authors received no external funding for this study.

Conflicts of Interests:
The authors have declared that no competing interests exist.

Data Sharing Statement:
The data that support the findings of this study are available on request from the corresponding author. The data is not publicly available due to privacy or ethical restrictions.

Received: February 06, 2025
1st Revision: June 02, 2026
2nd Revision: June 10, 2026
Accepted: June 13, 2026

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Original Article

Short-Term Clinical Outcomes of Kasai Porto-Enterostomy for Biliary Atresia: A Seven-Year Single-Center Experience

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https://doi.org/10.52206/jsmc.2026.16.3.1219

Abstract

Background: Biliary atresia is a rare but progressive fibro-obliterative cholangiopathy of infancy and represents the leading surgically correctable cause of neonatal cholestasis. Untreated disease progresses rapidly to biliary cirrhosis, liver failure, and death. Kasai Porto-Enterostomy (KPE) remains the primary surgical treatment for restoring bile flow and delaying or preventing liver transplantation. However, postoperative outcomes vary considerably among institutions, and local evidence from Pakistan remains limited.

Objective: To evaluate the clinical characteristics, perioperative complications, and short-term outcomes of infants undergoing Kasai Porto-Enterostomy for biliary atresia at a tertiary care referral center.

Methodology: This retrospective observational study was conducted in the Department of Pediatric Surgery, Saidu Group of Teaching Hospitals, Swat, Pakistan. Medical records of infants who underwent Kasai Porto-Enterostomy between January 2018 and January 2025 were reviewed. Patients undergoing surgery before 12 weeks of age and completing six months of follow-up were included. Demographic, clinical, laboratory, radiological, operative, histopathological, and follow-up data were extracted from hospital records.

Results: Thirty-six infants underwent Kasai Porto-Enterostomy during the study period. Two patients died during the immediate postoperative period, and one patient was lost to follow-up, leaving 33 patients for six-month outcome analysis. The mean age at surgery was 8.85 ± 1.00 weeks, and 54.5% of patients were female. Mean serum bilirubin decreased significantly from 5.89 ± 1.72 mg/dL preoperatively to 1.52 ± 0.24 mg/dL after surgery ($p < 0.001$). Clinical improvement with clearance of jaundice was observed in 78.8% of patients. Postoperative complications included bile leakage (21.2%), anastomotic leakage (12.1%), postoperative sepsis (9.1%), and surgical site infection (6.1%). Seven patients (21.2%) experienced postoperative cholangitis. No statistically significant association was observed between preoperative hepatic fibrosis and postoperative cholangitis ($p = 0.341$).

Conclusion: Kasai Porto-Enterostomy achieved significant short-term improvement in biliary drainage and clinical recovery in most infants with biliary atresia. Despite favorable biochemical and clinical outcomes, postoperative complications remained common, emphasizing the importance of meticulous perioperative care and structured postoperative follow-up. Larger prospective multicenter studies with long-term follow-up are required to evaluate native liver survival and identify factors influencing long-term outcomes.

Keywords: Biliary Atresia, Cholangitis, Kasai Porto-Enterostomy, Liver Fibrosis, Neonatal Cholestasis, Pediatric Surgery.

Introduction

Biliary atresia is a rare, progressive fibro-obliterative cholangiopathy affecting both the intrahepatic and extrahepatic biliary tree during early infancy. The disease initially presents with persistent neonatal cholestasis and progressive obstruction of bile flow, ultimately resulting in biliary cirrhosis, portal hypertension, liver failure, and death if left untreated [1]. Although its precise etiology remains uncertain, current evidence suggests that genetic susceptibility, immune dysregulation, viral infections, and environmental factors may contribute to disease pathogenesis [1].

The reported incidence of biliary atresia varies geographically, with the highest rates observed in Asian countries, affecting approximately one in every 5,000 to 20,000 live births [2]. In Pakistan, biliary atresia represents a significant cause of neonatal cholestatic jaundice. Azhar et al. reported that biliary atresia accounted for 26.7% of neonates presenting with prolonged jaundice at a tertiary care hospital, emphasizing the need for early diagnosis and prompt referral for surgical management [3].

Surgical intervention remains the cornerstone of treatment for biliary atresia. Kasai Porto-Enterostomy (KPE) is considered the standard first-line surgical procedure and aims to restore bile drainage by excising the fibrotic extrahepatic biliary remnant and creating a Roux-en-Y portoenterostomy [4]. Successful restoration of bile flow delays progression of liver disease, prolongs native liver survival, and reduces the immediate need for liver transplantation. Nevertheless, biliary atresia remains the most common indication for pediatric liver transplantation worldwide, accounting for nearly half of all liver transplants performed in children [5]. Contemporary studies report an approximate five-year native liver survival rate of 50% following successful Kasai Porto-Enterostomy [6].

The outcome of Kasai Porto-Enterostomy depends on several factors, including age at surgery, degree of hepatic fibrosis, postoperative biliary drainage, nutritional status, and the occurrence of postoperative complications [7]. Early diagnosis and timely surgical intervention have consistently been associated with improved clearance of jaundice and prolonged native liver survival. However, postoperative complications remain frequent, with reported rates ranging from 29% to 82%, including bile leakage, cholangitis, anastomotic complications, persistent jaundice, and progressive liver dysfunction [8]. Failure to establish adequate biliary drainage results in persistent cholestasis and remains one of the strongest predictors of early liver transplantation [9,10]. Among postoperative complications, cholangitis is particularly important because recurrent episodes accelerate hepatic fibrosis and significantly reduce native liver survival. A recent systematic review reported postoperative cholangitis in approximately 40%–93% of patients, most commonly during the first two years after surgery [11].

Although Kasai Porto-Enterostomy has become the standard initial treatment worldwide, reported outcomes differ substantially among institutions because of variations in referral patterns, timing of surgery, perioperative management, surgical expertise, and postoperative follow-up protocols. Published evidence from Pakistan remains scarce, particularly regarding short-term postoperative outcomes following Kasai Porto-Enterostomy. Consequently, locally generated data are required to better understand institutional outcomes, identify procedure-related complications, and guide quality improvement initiatives.

Therefore, the present study aimed to evaluate the demographic and clinical characteristics, perioperative complications, and short-term clinical and biochemical outcomes of infants undergoing Kasai Porto-Enterostomy for biliary atresia at a tertiary care teaching hospital. The findings are expected to contribute to the limited regional literature and provide evidence to optimize perioperative management and postoperative surveillance of children undergoing this procedure.

Methodology

Study Design

This retrospective observational study was conducted to evaluate the clinical profile, perioperative complications, and short-term outcomes of infants with biliary atresia who underwent Kasai Porto-Enterostomy (KPE). Medical records of eligible patients treated over a seven-year period, from January 2018 to January 2025, were reviewed. The study followed the Strengthening the Reporting of Observational Studies in Epidemiology (STROBE) recommendations for reporting observational research.

Ethics

Ethical approval was obtained from the Institutional Ethical Review Board of Saidu Group of Teaching Hospitals before commencement of the study. Owing to the retrospective nature of the study and the use of anonymized hospital records, the requirement for individual informed consent was waived by the Ethics Committee. Patient confidentiality was maintained throughout the study by assigning unique study identification numbers and removing all personal identifiers before data analysis.

Setting

The study was conducted in the Department of Pediatric Surgery, Saidu Group of Teaching Hospitals, Swat, Pakistan, which is a tertiary care referral center providing specialized pediatric surgical services for Khyber Pakhtunkhwa and surrounding regions. The institution routinely manages neonates and infants with congenital hepatobiliary disorders, including biliary atresia. Surgical procedures were performed by consultant pediatric surgeons according to institutional protocols, and all patients underwent standardized postoperative follow-up in the outpatient department.

Participants

Hospital records of all infants diagnosed with biliary atresia who underwent Kasai Porto-Enterostomy during the study period were reviewed. Diagnosis was established using clinical assessment, biochemical investigations, abdominal ultrasonography, intraoperative findings, and histopathological examination of liver biopsy specimens. Patients were eligible if they fulfilled all of the following criteria: (1) confirmed diagnosis of biliary atresia; (2) underwent Kasai Porto-Enterostomy before 12 weeks of age; and (3) completed at least six months of postoperative follow-up. Patients older than 12 weeks at surgery, those explored for suspected biliary atresia without undergoing Kasai Porto-Enterostomy, patients managed by alternative surgical procedures, and those with unavailable or incomplete follow-up records were excluded from the primary outcome analysis. During the study period, 36 infants underwent Kasai Porto-Enterostomy. Two patients died during the immediate postoperative period because of sepsis, and one patient was completely lost to follow-up. To minimize survival bias, the two postoperative deaths were included in the analysis of perioperative complications but excluded from six-month outcome analyses. The remaining 33 patients constituted the final study cohort for assessment of postoperative outcomes.

Variables

The primary outcome variable was the short-term outcome following Kasai Porto-Enterostomy assessed at six months after surgery. Successful outcome was defined by biochemical and clinical improvement, including reduction of serum bilirubin levels, restoration of normal stool pigmentation, resolution of jaundice, improvement in nutritional status, and absence of persistent cholestasis. Secondary outcome variables included postoperative complications such as bile leakage, anastomotic leakage, surgical site infection, postoperative sepsis, and cholangitis. Baseline explanatory variables included age at surgery, sex, preoperative gallbladder status on ultrasonography, preoperative serum bilirubin concentration, histopathological evidence of hepatic fibrosis, and postoperative serum bilirubin levels. For uniformity, biliary atresia was defined as a progressive fibro-obliterative cholangiopathy involving the extrahepatic biliary system causing obstruction of bile flow and progressive hepatic fibrosis. Kasai Porto-Enterostomy was defined as surgical excision of the obliterated extrahepatic biliary remnant followed by Roux-en-Y portoenterostomy to restore bile drainage. Postoperative cholangitis was defined as fever (>38.5°C) associated with recurrent jaundice, acholic stools, abdominal distension, or biochemical evidence of cholestasis, with or without microbiological confirmation. Operative morbidity included bile leak, anastomotic dehiscence, surgical site infection, postoperative sepsis, and portal fibrosis identified on histopathology.

Data sources/measurement

Clinical data were extracted independently from hospital admission registers, inpatient medical records, operative notes, anesthesia records, laboratory databases, radiology reports, histopathology reports, discharge summaries, and outpatient follow-up records using a standardized electronic data collection form designed in Google Forms. Data were subsequently exported into Microsoft Excel before statistical analysis. Preoperative ultrasonography was reviewed to determine gallbladder visualization. Serum total bilirubin values recorded before surgery and at the scheduled postoperative follow-up were retrieved from the hospital laboratory information system. Histopathological examination of ultrasound-guided liver biopsy specimens was reviewed to determine the presence or absence of hepatic fibrosis and ductular proliferation consistent with biliary atresia. Postoperative complications occurring during hospital admission were identified from daily progress notes, nursing records, operative re-intervention notes, microbiological reports, and discharge documentation. Six-month postoperative outcomes were obtained from outpatient clinic follow-up records.

Bias

Several measures were implemented to reduce potential sources of bias inherent to retrospective studies. Uniform eligibility criteria were applied to all patients. Data were extracted using a predefined standardized data collection form to minimize information bias. Multiple hospital records were cross-checked whenever discrepancies were identified. Objective laboratory parameters and documented clinical findings were used instead of subjective assessment wherever possible. Immediate postoperative deaths were retained in the perioperative complication analysis to reduce survival bias, while patients with completely unavailable follow-up data were excluded only from analyses requiring six-month outcome assessment.

Study size

Because biliary atresia is an uncommon congenital disorder, all eligible patients undergoing Kasai Porto-Enterostomy during the seven-year study period were included. Therefore, no formal sample size calculation was performed, and total population sampling was adopted to maximize statistical precision and minimize selection bias.

Quantitative variables

Continuous variables included age at surgery and serum bilirubin concentrations before and after surgery. These variables were summarized using means and standard deviations after assessment of their distribution. Categorical variables included sex, gallbladder visualization, histopathological fibrosis, postoperative complications, cholangitis, and clinical improvement, which were summarized using frequencies and percentages.

Statistical methods

Data were entered into Microsoft Excel, verified for completeness and consistency, and analyzed using Statistical Package for the Social Sciences (SPSS) version 26. Continuous variables were expressed as mean ± standard deviation (SD), whereas categorical variables were summarized as frequencies and percentages. The paired-samples t-test was used to compare mean preoperative and postoperative serum bilirubin concentrations. Fisher's exact test was applied to evaluate the association between preoperative hepatic fibrosis and postoperative cholangitis because of the relatively small sample size and low expected cell counts. All statistical tests were two-tailed, and a p-value ≤0.05 was considered statistically significant.

Results

Participants

During the study period from January 2018 to January 2025, 36 infants underwent Kasai Porto-Enterostomy for biliary atresia at the study center. Two patients died during the immediate postoperative period because of sepsis, while one patient was lost to follow-up because postoperative records were unavailable. To minimize survival bias, the two perioperative deaths were included in the analysis of postoperative complications but excluded from the six-month outcome assessment. Consequently, 33 patients were included in the final analysis of clinical and biochemical outcomes.

Descriptive data

The baseline demographic and preoperative characteristics of the study population are summarized in Table 1. Of the 33 patients included in the final analysis, 18 (54.5%) were female and 15 (45.5%) were male. The age at surgery ranged from 7 to 12 weeks, with a mean age of 8.85 ± 1.00 weeks. Preoperative ultrasonography demonstrated a rudimentary, non-functional gallbladder in 30 patients (90.9%), whereas the gallbladder was not visualized in three patients (9.1%). Preoperative serum bilirubin levels ranged from 3.0 to 9.0 mg/dL, with a mean concentration of 5.89 ± 1.72 mg/dL. Histopathological examination of liver biopsy specimens demonstrated ductular proliferation compatible with biliary atresia in all patients. Hepatic fibrosis was identified in 24 patients (72.7%), while nine patients (27.3%) had no histological evidence of fibrosis (Table 1).

Outcome data

Postoperative biochemical outcomes are presented in Table 2. At postoperative follow-up, serum bilirubin levels decreased significantly, with a mean postoperative concentration of 1.52 ± 0.24 mg/dL (range: 0.75–2.43 mg/dL). Comparison of preoperative and postoperative bilirubin concentrations using the paired t-test demonstrated a statistically significant reduction following Kasai Porto-Enterostomy ($p < 0.001$). Postoperative complications occurred in 16 of 33 patients (48.5%), whereas 17 patients (51.5%) had an uncomplicated postoperative course. The most frequent complication was bile leakage, occurring in seven patients (21.2%), followed by anastomotic leakage in four patients (12.1%), postoperative sepsis in three patients (9.1%), and surgical site infection in two patients (6.1%) (Table 2). When the two immediate postoperative deaths were included, the overall perioperative adverse event rate was 50.0% (18/36). At six months after surgery, 26 patients (78.8%) achieved clinical improvement with clearance of jaundice, whereas seven patients (21.2%) had persistent jaundice. During follow-up, seven patients (21.2%) experienced a single documented episode of postoperative cholangitis (Table 2).

Main results

The relationship between preoperative hepatic fibrosis and postoperative cholangitis is presented in Table 3. Among patients who developed postoperative cholangitis, five (71.4%) had histological evidence of hepatic fibrosis and two (28.6%) had no fibrosis. Among patients without cholangitis, 19 (73.1%) had fibrosis and seven (26.9%) had no fibrosis. Fisher's exact test demonstrated no statistically significant association between preoperative hepatic fibrosis and the occurrence of postoperative cholangitis ($p = 0.341$). Overall, Kasai Porto-Enterostomy was associated with a significant reduction in postoperative serum bilirubin concentrations ($p < 0.001$). Clinical improvement was observed in 78.8% of patients at six months, while postoperative complications were documented in 48.5% of patients.

Table 1: Baseline Demographic and Preoperative Characteristics of Patients Undergoing Kasai Porto-Enterostomy (n = 33)	
Variables	**N(%)**
Age at surgery (weeks); Mean ± SD	8.85 ± 1.00
Sex	
Male	15 (45.5%)
Female	18 (54.5%)
Preoperative gallbladder status	
Visualized as rudimentary/non-functional	30 (90.9%)
Non-visualized	3 (9.1%)
Preoperative serum bilirubin (mg/dL); Mean ± SD	5.89 ± 1.72
Histopathology	
Fibrosis	24 (72.7%)
No fibrosis	9 (27.3%)

Table 2: Postoperative Outcomes Following Kasai Porto-Enterostomy (n = 33)	
Variables	**N(%)**
Postoperative serum bilirubin (mg/dL); Mean ± SD	1.52 ± 0.24
Clinical improvement at 6 months	
Improved (clearance of jaundice)	26 (78.8%)
Persistent jaundice	7 (21.2%)
Postoperative complications	
No complications	17 (51.5%)
Bile leak	7 (21.2%)
Anastomotic leak	4 (12.1%)
Sepsis	3 (9.1%)
Surgical site infection	2 (6.1%)

Table 3: Association Between Preoperative Hepatic Fibrosis and Postoperative Cholangitis (n = 33)			
Histopathology	**Cholangitis (n = 7)**	**No Cholangitis (n = 26)**	**p-value**
Fibrosis	5 (71.4%)	19 (73.1%)	0.341
No fibrosis	2 (28.6%)	7 (26.9%)	

*p<0.05 is considered statistically significant

Discussion

Kasai Porto-Enterostomy remains the primary surgical treatment for biliary atresia and aims to establish biliary drainage, delay progression of chronic liver disease, and prolong native liver survival. The present study evaluated the short-term outcomes of Kasai Porto-Enterostomy over a seven-year period at a tertiary care pediatric surgical center. The findings demonstrated significant postoperative improvement in serum bilirubin levels, with nearly four-fifths of patients achieving clinical improvement within six months. However, postoperative complications, particularly bile leakage and cholangitis, remained relatively common.

The timing of surgery is considered one of the most important determinants of outcome following Kasai Porto-Enterostomy. Earlier intervention has consistently been associated with improved biliary drainage and better long-term native liver survival [12,13]. In the present study, the mean age at surgery was 8.85 ± 1.00 weeks, indicating that most infants underwent surgery within the generally recommended therapeutic window. Our findings are consistent with those reported by Andrade et al., who observed that improvements in early diagnosis and referral have progressively reduced the age at surgery compared with historical cohorts, thereby improving postoperative outcomes [14]. Although our study did not specifically evaluate the influence of operative timing on outcomes because of the relatively narrow age range of participants, the favorable rate of jaundice clearance observed in our cohort may partly reflect timely surgical intervention.

Successful clearance of jaundice following Kasai Porto-Enterostomy is widely recognized as an early surrogate marker of adequate biliary drainage and is strongly

associated with improved native liver survival [10]. In the present study, 78.8% of patients demonstrated clinical improvement with clearance of jaundice within six months after surgery. This rate is higher than the 62.9% reported by Liu et al. [15] and substantially exceeds the 20.9% reported by Wickramasekara et al. [16]. Differences among studies may be explained by variation in patient selection, disease severity at presentation, timing of surgery, duration of follow-up, institutional surgical expertise, and postoperative management protocols. Nevertheless, our findings fall within the upper range of jaundice clearance rates reported in the international literature, where successful biliary drainage has generally been observed in approximately 50–70% of patients after Kasai Porto-Enterostomy [17].

The influence of age at surgery on postoperative jaundice clearance remains controversial. Japanese registry data demonstrated superior outcomes with earlier surgical intervention [18], whereas studies from the United States reported no significant relationship between operative age and jaundice clearance [19]. Conversely, a retrospective Chinese study suggested that extremely early surgery may not always translate into improved postoperative outcomes because disease severity and hepatic pathology also influence prognosis [20]. Owing to the limited variation in operative age within our study population (7–12 weeks), meaningful subgroup analysis could not be performed. Consequently, the present study neither confirms nor refutes the influence of operative timing on postoperative jaundice clearance.

Postoperative cholangitis is one of the most frequently encountered complications after Kasai Porto-Enterostomy and remains an important determinant of long-term native liver survival [11]. In the present study, postoperative cholangitis occurred in 21.2% of patients, which is comparable to previously reported rates ranging from 20% to 35% [21]. The occurrence of cholangitis has been associated with several clinical factors, including impaired biliary drainage, malnutrition, bacterial translocation, and advanced hepatic fibrosis [22]. Although hepatic fibrosis was more frequently observed among patients who developed cholangitis, statistical analysis demonstrated no significant association between fibrosis and postoperative cholangitis ($p = 0.341$). This lack of significance is likely attributable to the relatively small sample size rather than the absence of a true biological relationship. Previous studies have similarly produced inconsistent findings regarding predictors of postoperative cholangitis [23,24].

The incidence of postoperative bile leakage (21.2%) and anastomotic leakage (12.1%) in our study was higher than rates commonly reported in the literature, where these complications generally occur in fewer than 10% of patients [24]. Several factors may explain this observation. First, our institution serves as a tertiary referral center, and many infants presented with advanced disease characterized by hepatic fibrosis, cholestasis, hypoalbuminemia, and poor nutritional status, all of which may impair tissue healing and increase the risk of postoperative complications. Second, the relatively small sample size may have amplified the apparent frequency of adverse events. Finally, variations in surgical technique, perioperative care, and complication reporting among institutions may also contribute to differences between published studies.

Despite these postoperative complications, Kasai Porto-Enterostomy achieved a significant reduction in serum bilirubin concentrations following surgery ($p < 0.001$), indicating successful restoration of biliary drainage in the majority of patients. These findings support the continued role of Kasai Porto-Enterostomy as the preferred initial surgical treatment for biliary atresia, particularly when performed early in appropriately selected patients and followed by comprehensive postoperative surveillance.

Limitations

Several limitations should be considered when interpreting the findings of this study. First, this was a retrospective single-center observational study, making it susceptible to selection bias, incomplete documentation, and information bias inherent to medical record reviews. Second, the relatively small sample size, reflecting the rarity of biliary atresia, limited the statistical power to detect associations between clinical variables and postoperative outcomes, particularly in subgroup analyses. Third, the study evaluated only short-term outcomes over a six-month follow-up period. Consequently, important long-term endpoints such as native liver survival, transplant-free survival, portal hypertension, growth and nutritional status, quality of life, and overall survival could not be assessed. Fourth, although postoperative bilirubin reduction was used as an objective indicator of successful biliary drainage, additional prognostic biomarkers, liver function parameters, and standardized nutritional assessments were not consistently available because of the retrospective design.

Finally, the study was conducted at a single tertiary referral center, where patients frequently presented with advanced disease and hepatic fibrosis. Therefore, the findings may not be fully generalizable to other institutions, community hospitals, or healthcare systems with different referral pathways, patient characteristics, or surgical expertise. Prospective multicenter studies with larger sample sizes and extended follow-up are required

to validate these findings and identify independent predictors of long-term outcomes after Kasai Porto-Enterostomy.

Conclusion

Kasai Porto-Enterostomy provided favorable short-term clinical and biochemical outcomes in infants with biliary atresia, with significant postoperative reduction in serum bilirubin levels and clinical improvement observed in nearly four-fifths of patients within six months of surgery. Although postoperative complications, including bile leakage, anastomotic leakage, and cholangitis, were relatively common, most were successfully managed during the postoperative period.

Authors' Contributions:

All authors meet the ICMJE criteria for authorship.

Author 1: Substantial contribution to conception and design, drafting the manuscript, final approval, agreed to be accountable for all the work.
Author 2: Substantial contribution to conception and design, data acquisition, drafting the manuscript, final approval, agreed to be accountable for all the work.
Author 3: Analysis and interpretation of data, drafting the manuscript, final approval, agreed to be accountable for all the work.
Author 4: Analysis and interpretation of data, critical revision, final approval, agreed to be accountable for all the work.
Author 5: Substantial contribution to conception and design, drafting the manuscript, final approval, agreed to be accountable for all the work.
Author 6: Substantial contribution to conception and design, data acquisition, drafting the manuscript, final approval, agreed to be accountable for all the work.

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