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Predictors of Short-Term Outcome of Kasai Portoenterostomy for Biliary Atresia in Infants: a Single-Center Study

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Conflict of Interest
The authors have no financial conflicts of interest.

ABSTRACT

Purpose: The outcome predictors of Kasai portoenterostomy (KPE) for biliary atresia (BA) are controversial. This study aimed to identify possible short-term outcome predictors of KPE for BA in infants.
Methods: This retrospective study included infants with BA who underwent KPE between January 2015 and December 2017 and were followed up for at least 6 months after surgery at the Pediatric Hepatology Unit, Cairo University Pediatric Hospital, Egypt. The short-term outcome was jaundice clearance within 6 months following surgery. All data were compared between the jaundice free group and those with persistent jaundice to identify the predictors of jaundice clearance.
Results: The study included 75 infants. The mean age at the time of surgery was 82.43±22.77 days (range, 37–150 days), and 28 (37.3%) infants cleared their jaundice within 6 months postoperative. Age at surgery did not significantly affect the outcome (p=0.518). Infants with persistent jaundice had significantly higher pre-operative levels of aspartate aminotransferase (AST) than those who were jaundice free (p=0.041). Receiver operating characteristic curve analysis showed that preoperative AST ≤180 IU/L was predictive of a successful KPE, with sensitivity 74.5% and specificity 60.7%. Infants with bile plugs in liver biopsy had a 6-fold higher risk of persistent jaundice than those without bile plugs (95% confidence interval: 1.59–20.75, p=0.008).
Conclusion: Jaundice clearance after KPE for BA can be predicted using preoperative AST and presence of bile plugs in liver biopsy.

Keywords: Biliary atresia; Portoenterostomy, hepatic; Outcome

INTRODUCTION

Biliary atresia (BA) is a rare disease that affects an estimated 1 in 8,000–18,000 live births [1]. Nearly half of the affected children will eventually need liver transplantation (LT) within the first 2 years of life. Furthermore 20%–30% will need LT in childhood and adolescence [2].

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Kasai portoenterostomy (KPE) is the best treatment for BA. Total extraction of the hilar fibrosis and subsequent biliary-enteric anastomosis results in a favorable outcome with total restitution of the biliary drainage and normalization of total bilirubin levels in about two-third of cases [3].

There are many factors that can influence the success rate of KPE including the patient's age at the time of surgery, presence of cirrhosis, surgeon's experience, postoperative cholangitis, and unknown genetic factors [4]. Age alone cannot predict the outcome [5].

Therefore, we conducted this single-center study to determine the factors predicting the short-term outcome of KPE.

MATERIALS AND METHODS

This retrospective cohort study included infants with BA who underwent KPE between January 2015 and December 2017. The study was conducted at the Pediatric Hepatology Unit, Cairo University Pediatric Hospital, Egypt. All patients with BA who underwent KPE in our hospital during these 3 years and were followed for at least 6 months after surgery were included in the study. Patients with insufficient data in their files and those who were lost to follow-up were excluded.

The following data were retrieved from the patients' files:

- Gestational age
- Gender
- Age at referral
- Age at operation
- Presence of congenital malformations (congenital heart disease, biliary atresia splenic malformation [BASM])
- Growth parameters at presentation
- Liver function tests at presentation and 6 months after surgery: total and direct serum bilirubin, alanine aminotransferase (ALT), aspartate aminotransferase (AST), alkaline phosphatase (ALP), gamma glutamyl transpeptidase (GGT), serum albumin, and international normalised ratio (INR)
- Preoperative AST/ALT ratio was calculated
- Histopathological findings of preoperative liver biopsy
- Preoperative abdominal ultrasonography
- Development of cholangitis (fever, clay colored stool, leukocytosis, hyperbilirubinemia, and bacteremia) or portal hypertension (bleeding esophageal varices), and liver cell failure (ascites, coagulopathy) in the 6 months following KPE

All patients received postoperative intravenous antibiotics. Since March 2016, patients were randomized into 2 arms: one arm received oral prednisone starting from 5th postoperative day at a dose of 5 mg/kg/day that was tapered by 1 mg/kg every 5 days and discontinued after 25 days, whereas the other arm served as controls. Ursodeoxycholic acid (dose 10–15 mg/kg/day) was administered only after re-coloration of stool. All patients received fat soluble vitamins. Prophylactic trimethoprim/sulfamethoxazole was prescribed to all infants after surgery and was continued for 12 months postoperative.

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The short-term outcome was clearance of jaundice within 6 months postoperative. Accordingly, patients were classified into jaundice-free (JF) group (total serum bilirubin <2 mg/dL) and persistent jaundice (PJ) group (total serum bilirubin >2 mg/dL).

All parents/guardians signed an informed consent, and the study was reviewed and approved by the Cairo University Research ethics committee (REC) (N-I-111015-2018) and was performed in accordance with the Declaration of Helsinki.

Statistical analyses

The median values along with the interquartile range of preoperative and 6 months postoperative liver function tests were calculated. According to the central limit theorem, as there were 30 records in each group, the *p*-value was calculated using the Student t-test. Correlation between various variables was analyzed using Spearman rank correlation equation. For comparing categorical data, Chi-square (χ^2) test was performed, and Exact test was used when the expected frequency was <5. Accuracy was represented using the terms sensitivity and specificity. Receiver operating characteristic (ROC) analysis was used to determine the optimum cut-off value of AST that predicted poor outcome. Two-sided *p*-values <0.05 were considered statistically significant. All statistical analyses were performed using IBM SPSS Statistics for Windows, Version 22.0 (IBM Corp., Armonk, NY, USA).

RESULTS

This study included 75 children diagnosed with BA who underwent KPE and were followed up for 6 months after surgery. They were 45 (60.0%) boys and 30 (40.0%) girls. Consanguineous marriage was present in 27 families (36.0%). Two (2.7%) were preterm babies. Two (2.7%) infants had BASM, and one of them had situs inversus totalis. Four (5.3%) had congenital heart disease. The bodyweight, height, and skull circumference were below the 3rd percentile for age in 1%, 3%, and 7% respectively. The mean age at referral and time of surgery was 67±24 and 82±23 days, respectively, with the youngest and oldest being 30 and 150 days old, respectively.

The gall bladder was visualized preoperatively by abdominal ultrasonography in 13 (17.3%) infants, and associated cystic lesion was present in 4 (5.3%). Splenomegaly was found in 32 (42.7%) infants.

The JF group included 28 (37.3%) infants who cleared their jaundice within 6 months post KPE, while there were 47 (62.7%) infants in the PJ group. Data of both groups were compared to identify factors associated with clearance or persistence of jaundice. The demographic and clinical characteristics are shown in **Table 1**. Age at surgery did not significantly affect the outcome (*p*=0.510).

Preoperative liver biopsy was available for 73 (97.3%) infants. All infants had bile duct proliferation and hepatocellular degeneration. Impending cirrhosis was present in only 2 (2.7%) infants who were 74 and 86 days old at surgery; the former was JF 6 months postoperative. Bile plugs were significantly more common in the PJ group than the JF group (*p*=0.013). Multivariate analysis showed that the presence of bile plugs was an independent risk factor for PJ (relative risk [RR]: 6, 95% confidence interval [CI]: 1.59–20.75, *p*=0.008). Infants with bile plugs had about 6-fold higher risk of PJ at 6 months postoperative than those without bile plugs. There was a weak but significant correlation between the degree of

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portal fibrosis and the age of KPE (Spearman's correlation coefficient 0.237; p=0.047). Other histological findings did not significantly affect the outcome (**Table 2**).

Liver function tests were compared both preoperative (**Table 3**) and 6 months postoperative (**Table 4**). Only preoperative AST was significantly higher in the PJ group than in the JF group (p=0.041). Multivariate analysis showed that AST was an independent risk factor for PJ (RR: 1, 95% CI: 1–1.01, p=0.040). ROC curve analysis for AST was performed where the area under

Table 1. Comparison between demographic and clinical characteristics of both groups of infants with biliary atresia

Variable	Jaundice-free at 6 months (n=28)	Persistent jaundice at 6 months (n=47)	Total (n=75)	*p*-value
Age of referral (d)	67±24	68±24	67±24	0.860
Age at operation (d)	80±17	84±26	82±23	0.510
Gender				0.630
Male	18 (64.3)	27 (57.3)	45 (60.0)	
Female	10 (35.7)	20 (42.6)	30 (40.0)	
Prematurity	1 (3.6)	1 (2.1)	2 (2.7)	NA
Weight-for age <3rd percentile	1 (3.6)	0 (0)	1 (1.3)	NA
Length-for age <3rd percentile	0 (0)	2 (4.3)	2 (2.7)	NA
Head circumference for age <3rd percentile	2 (7.1)	3 (6.4)	5 (6.7)	1
Splenomegaly	11 (39.3)	21 (45.0)	32 (42.7)	0.810
BASM	1 (3.6)	1 (2.1)	2 (2.7)	NA
Congenital heart disease	1 (3.6)	3 (6.1)	4 (5.3)	1

Values are presented as mean±standard deviation or number (%).
BASM: biliary atresia splenic malformation, NA: not applicable.

Table 2. Comparison between histopathological findings in preoperative liver biopsy in both groups of infants with biliary atresia (n=73)

Histopathologic findings	Jaundice-free at 6 months (n=27)	Persistent jaundice at 6 months (n=46)	*p*-value
Distorted lobular architecture	21 (77.8)	39 (84.8)	0.220
Ductal cholestasis	8 (29.6)	13 (28.3)	0.900
Bile plugs	12 (44.4)	29 (63.0)	0.013
Degree of portal fibrosis			0.790
Mild	5 (18.5)	7 (15.2)	
Moderate	14 (51.9)	27 (58.7)	
Severe	5 (18.5)	12 (26.1)	
Fibrous septa	11 (40.7)	21 (45.7)	0.180
Porto-portal bridging fibrosis	13 (48.1)	22 (47.8)	0.330
Hepatocellular degeneration	25 (92.6)	44 (97.9)	NA
Giant cells	13 (48.1)	18 (39.1)	0.450
Hyperplastic Von Kupffer cells	0 (0)	5 (10.9)	0.070

Values are presented as number (%).
NA: not applicable.

Table 3. Comparison between preoperative liver function tests in both groups of infants with biliary atresia

Preoperative laboratory findings	Jaundice-free at 6 months (n=28)	Persistent jaundice at 6 months (n=47)	*p*-value
TB (mg/dL)	9.8 (2.6)	10.5 (3.5)	0.470
DB (mg/dL)	5.9 (2.3)	7 (2.9)	0.080
ALT (IU/L)	105 (136)	120 (106)	0.540
AST (IU/L)	174.5 (137)	222 (151)	0.040
AST/ALT ratio	1.7 (0.7)	1.8 (0.8)	0.390
GGT (IU/L)	846.5 (842)	802 (682)	0.450
ALP (IU/L)	683 (495)	611 (364)	0.710
Albumin (g/dL)	3.5 (0.9)	3.3 (1.0)	0.190
INR	1 (0)	1 (0)	0.430

Values are expressed as median (interquartile range).
TB: total bilirubin, DB: direct bilirubin, ALT: alanine aminotransferase, AST: aspartate aminotransferase, GGT: gamma glutamyl transpeptidase, ALP: alkaline phosphatase, INR: international normalized ratio.

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ROC curve (AUC) of AST was 0.64 (95% CI 0.51–0.77) (**Fig. 1**). With this analysis, the optimal cut-off value for AST was determined as 180 IU/L. The calculated sensitivity and specificity based on this cut-off value were 74.5% and 60.7%, respectively. Preoperative AST/ALT ratio was not significant between the two groups (**Table 3**). Postoperative AST was significantly lower in the JF group than in the PJ group (p=0.001). INR and serum albumin were significantly different between the two groups (p=0.002 and p<0.0001, respectively) (**Table 4**).

Out of 31 (41.3%) infants who received adjuvant steroids, 9 (29.0%) cleared their jaundice within 6 months but 22 (71.0%) did not. Therefore, the use of steroids did not significantly affect the outcome (p=0.210).

During the follow-up period, 21 (28.0%) infants developed cholangitis (**Table 5**). Although cholangitis occurred more frequently in the PJ group than in the JF group (34% vs. 17.9%),

Table 4. Comparison between liver function tests in both groups of infants with biliary atresia 6 months after Kasai portoenterostomy

Postoperative laboratory findings	Jaundice-free at 6 months (n=28)	Persistent jaundice at 6 months (n=47)	*p*-value
ALT (IU/L)	65.5 (71)	101 (104)	0.090
AST (IU/L)	84 (71)	167 (163)	<0.001
ALP (IU/L)	497 (517)	602 (372)	0.740
GGT (IU/L)	419 (503)	414 (646)	0.410
Albumin (g/dL)	3.7 (0.8)	2.7 (0.7)	<0.001
INR	1 (0)	1.1 (0)	<0.001

All values are expressed as median (interquartile range).
ALT: alanine aminotransferase, AST: aspartate aminotransferase, ALP: alkaline phosphatase, T: gamma glutamyl transpeptidase, INR: international normalized ratio.

Table 5. Frequency of complications in both groups of infants with biliary atresia within 6 months after Kasai portoenterostomy

Complications	Jaundice-free at 6 months (n=28)	Persistent jaundice at 6 months (n=47)	Total (n=75)	*p*-value
Cholangitis	5 (17.9)	16 (34.0)	21 (28.0)	0.080
Coagulopathy	1 (3.6)	10 (21.3)	11 (14.7)	0.040
Ascites	0 (0)	19 (40.4)	19 (25.3)	<0.001
Hematemesis	2 (7.1)	4 (8.5)	6 (8.0)	0.810

Values are presented as number (%).

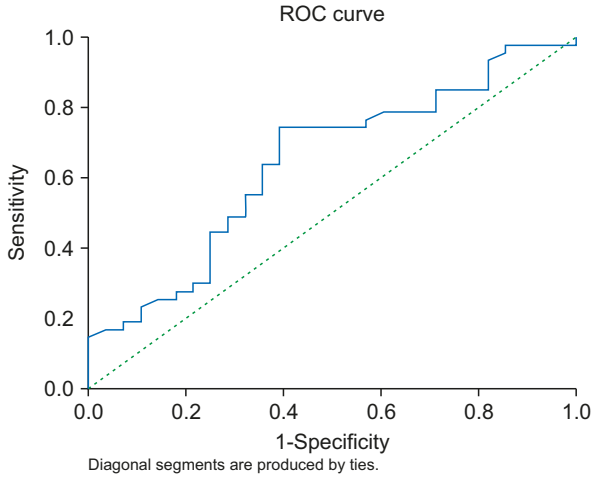

Fig. 1. Receiver operating characteristic curve for aspartate aminotransferase (AST).
ROC: receiver operating characteristic.

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the difference was not significant (p=0.080). During follow-up, worsening of coagulopathy and development of ascites was more common in the PJ group than in the JF group (21.3% vs. 3.6%, p=0.041 and 40.4% vs. 0%, p<0.0001, respectively). The frequency of KPE complications are shown in **Table 5**.

DISCUSSION

Despite the high birth rate in Egypt and the not uncommon presentation of cases with BA at our high volume tertiary referral center, this study is one of the few reports from Egypt that analyzed the underlying factors affecting the outcome of KPE. The aim of centralisation is to allow the best treatment for cases. A center doing at least 5 cases/year is recommended to achieve a better outcome [6] this was easily achieved in our study. Jaundice clearance was achieved in 37% of our study population, which is higher than that reported previously in a study from Egypt (27%) [7] but lower than that reported in other studies (55%) [8]. This low rate of jaundice clearance in our study may be attributed to the older age at referral in our cohort (67±24 days). However, in the previous study from Egypt [7], the mean age at surgery in the JF group was younger than in our study (70±12 days), which suggests that factors other than the age at operation can influence the outcome. KPE continues to be relevant because up to 38% of children can survive with their native livers for 30 years after KPE [9].

Age at Kasai portoenterostomy

Although the mean age at referral was late in our study, the mean age at surgery was 80±17 days. However, the latter did not significantly affect the outcome. A Canadian study, almost a decade ago, reported a median age at referral of 55 days [10], whereas a recent Brazilian study reported a median age of 60 days [11]. Some studies have shown that the high age at KPE did not predict a favorable outcome [12,13]. We do not suggest delaying the age at KPE, but we want to emphasize that the notion of imperative early surgery may not apply to all patients, and late presenting cases may still benefit from KPE. Our oldest case at operation who cleared their jaundice was 117 days old.

Davenport et al. [14] attributed that the discrepancies in the outcome are caused in part by the heterogeneous nature of the older infants with BA; presumably it is an acquired BA, in whom intrinsic parenchymal disease is not too advanced, unlike those in whom the diagnosis is delayed owing to failure of realizing the importance of persistence of jaundice. BA should not be thought as a single disease entity with a predictable natural history and stereotypical response [15]. Performing late KPE after 90 days of age has still a benefit delaying the need for LT and immunosuppression [16]. Chardot et al. [17] reported that the 5-year survival with native liver in infants older than 3 months at the time of KPE was 45% which justifies performing KPE after 3 months of age in selected cases without advanced liver disease. On the other hand, some studies showed that the outcome was best when infants were operated before 60 days of age [18,19] or before 72 days [20]. Nio et al. [21] noted that the jaundice disappearance rate was 100%, 69.8%, 57%, 41%, 7.7%, and 0% in infants operated at <30, 31–60, 61–90, 91–120, 121–150, and >150 days of age, respectively and concluded that the operative age had a significant impact on the short term jaundice disappearance rate but might not have a significant impact on the long term ultimate native liver survival rate if KPE was done before 4 months of age.

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The only contraindications for KPE are liver cell failure, portal hypertension, and uncorrectable coagulopathy which were not present in this study. These children have rapid decompensation in the postoperative period, resulting in a high morbidity and mortality [16].

Sex, prematurity, and anthropometric measurements

Sixty percent of our infants were boys, in contrast to other studies that described slight predominance of girls for BA [22,23]. The majority of our cases had normal anthropometric measurements. In a previous study the bodyweight was not significantly associated with high mortality [24], while another study reported that infants with bodyweight less than 3.5 kg had significantly higher mortality than those with bodyweight >3.5 kg [23]. The present study included 2 infants who were born prematurely; this small number did not enable us to draw an association between prematurity and outcome.

Syndromic presentation

We had only 2 patients with BASM, one of which was JF at the end of the study. Poor prognosis was reported in patients with polysplenia [17,23]. Only 4 infants had congenital heart disease, with no significance regarding jaundice clearance. The presence of other congenital anomalies was reported as one of the predictors of early failure after KPE [13].

Histopathology

We assumed that the histopathological features can predict the outcome. Bile plugs were a significant predictor of PJ. Other histologic findings including the presence of giant cells, distorted lobular architecture, or portal fibrosis had no clinical significance. In a Brazilian study, most patients had bile plugs (94%), ductular/ductal proliferation (94%), and fibrosis (84%) [24]. In a previous study, the histopathological examination was not discriminatory and could not be used as a predictor [14,12]. However, histological criteria predicted outcome in an earlier study by Azarow et al. [25]; the presence of syncitial giant cells, lobular inflammation, focal necrosis, and bridging necrosis were each associated with the failure of KPE (p<0.050). Impending cirrhosis was present in only 2 (3%) infants who were 74 and 86 days old at surgery, the former was JF at 6 months postoperative. However, in another study, the features of early cirrhosis were only found in patients who were more than 100 days of age at surgery; none of these infants had a successful outcome [26].

Liver function tests

In the current study, the preoperative serum level of AST was a good outcome predictor, and it was significantly higher in the PJ group than in the JF group. Moreover, AST 6 months postoperative was significantly lower in the JF group than in the PJ group. Other authors found that the preoperative AST was elevated significantly in all children with BA, postoperatively it declined significantly by 2-fold in the anicteric group [27].

A previous study has shown that the total and direct bilirubin levels reflect the degree of cholestasis, while ALT and AST levels reflect inflammation and damage of liver cells with future fibrosis. Serum total and direct bilirubin and AST and ALT were significantly higher in those who required LT or died than those who survived with their native liver [28]. They set the 2 months postoperative cut-off values for direct bilirubin and AST at 0.7 mg/dL and 94 IU/L, respectively. Infants below these cut-off values had better survival rates with their native liver than those above these cut-off values. AST level at 1 year postoperative can be a predictive variable for the quality of life and liver dysfunction in long term JF infants [29].

In the aforementioned study of Abdel-Aziz et al. [7] 2019, ALP had a good performance in predicting the outcome. A cut-off value of ALP at 532.5 U/L or less predicted a successful KPE with 75.9% sensitivity and 74.4% specificity [7]. A preoperative total bilirubin level >10 mg/dL was found to be a poor prognostic factor in a previous study [30].

Nevertheless, other studies reported that the biochemical profile of the patients helped in their pre- and postoperative assessment but were not significantly associated with a favorable outcome [31].

In our cohort, the use of steroids did not significantly affect the outcome. Other studies had similar results [32] or reported an overall non-significant increase in jaundice clearance at 6 months (49% vs. 59%) in the steroid group [33]. However, Dong et al. [34] found a significant difference in favor of the steroid group.

We observed that repeated cholangitis was not a significant risk factor for predicting failure of KPE. Postoperative cholangitis occurred in 30% infants, which is almost similar to 21% reported by Redkar et al. [18], and less than 53% reported by Al-Kawaz et al. [23] Other studies observed that the only significant prognostic variable predicting KPE outcome was the occurrence of recurrent cholangitis as bile drainage is compromised by bacteria and inflammation in the sclerotic bile ductules and cholestasis leading to further liver injury and fibrotic changes [13,30].

Moreover, gastrointestinal bleeding was present in only 8% which was lower than that reported by other studies (16% and 29%) [35,36].

Strengths and limitations

One of the strengths of our study is the adequate sample size from a single center. However, a limitation is the retrospective nature of the study.

Conclusions and recommendations

The presence of bile plugs was one of the key histopathological features that predicted failure of KPE. AST is a simple biochemical marker that should be consistently evaluated in patients. Elevated preoperative AST was a poor prognostic factor. We identified AST ≥180 IU/L as an optimal cut-off value for predicting poor outcome. In our study, the presence of histological evidence of impending cirrhosis did not prevent a 74-day-old infant from becoming JF. Predictors can be used objectively to discuss with the parents the outcome of KPE and their effect on LT. We advise that KPE should be offered to underprivileged late presenting cases between 90 and 120 days of age.

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