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World J Transplant 2021 September 18; 11(9): 356-409



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INDEXING/ABSTRACTING

The WJT is now abstracted and indexed in PubMed, PubMed Central, Scopus, China National Knowledge Infrastructure (CNKI), and Superstar Journals Database.

RESPONSIBLE EDITORS FOR THIS ISSUE

Production Editor: *Ying-Yi Yuan*, Production Department Director: *Yu-Jie Ma*, Editorial Office Director: *Jia-Ping Yan*.

NAME OF JOURNAL

World Journal of Transplantation

ISSN

ISSN 2220-3230 (online)

LAUNCH DATE

December 24, 2011

FREQUENCY

Monthly

EDITORS-IN-CHIEF

Maurizio Salvadori, Sami Akbulut, Vassilios Papalois

EDITORIAL BOARD MEMBERS

<https://www.wjnet.com/2220-3230/editorialboard.htm>

PUBLICATION DATE

September 18, 2021

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<https://www.wjnet.com/bpg/gerinfo/240>

PUBLICATION ETHICS

<https://www.wjnet.com/bpg/gerinfo/288>

PUBLICATION MISCONDUCT

<https://www.wjnet.com/bpg/gerinfo/208>

ARTICLE PROCESSING CHARGE

<https://www.wjnet.com/bpg/gerinfo/242>

STEPS FOR SUBMITTING MANUSCRIPTS

<https://www.wjnet.com/bpg/gerinfo/239>

ONLINE SUBMISSION

<https://www.f6publishing.com>



Management of biliary atresia: To transplant or not to transplant

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Author contributions: Kakos CD, Ziogas IA, Alexopoulos SP and Tsoulfas G conceived and designed the study, acquired, analyzed, and interpreted the data, drafted, and critically revised the manuscript, and approved the final version of the manuscript.

Conflict-of-interest statement: The authors declare no conflict of interests for this article.

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Manuscript source: Invited manuscript

Specialty type: Transplantation

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Abstract

Kasai procedure (KP) and liver transplantation (LT) represent the only therapeutic options for patients with biliary atresia (BA), the most common indication for LT in the pediatric population. However, KP represents by no means a radical option but rather a bridging one, as nearly all patients will finally require a liver graft. More and more experts in the field of transplant surgery propose that maybe it is time for a paradigm change in BA treatment and abandon KP as transplantation seems inevitable. Inadequacy of organs yet makes this option currently not feasible, so it seems useful to find ways to maximize the efficacy of KP. In previous decades, multiple studies tried to identify these factors which opt for better results, but in general, outcomes of KP have not improved to the level that was anticipated. This review provides the framework of conditions which favor native liver survival after KP and the ones which optimize a positive LT outcome. Strategies of transition of care at the right time are also presented, as transplantation plays a key role in the surgical treatment of BA. Future studies and further organization in the transplant field will allow for greater organ availability and better outcomes to be achieved for BA patients.

Key Words: Biliary atresia; Kasai procedure; Portoenterostomy; Native liver survival; Liver transplantation

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Country/Territory of origin: Greece**Peer-review report's scientific quality classification**

Grade A (Excellent): 0
 Grade B (Very good): 0
 Grade C (Good): C
 Grade D (Fair): 0
 Grade E (Poor): 0

Received: May 20, 2021**Peer-review started:** May 20, 2021**First decision:** June 17, 2021**Revised:** June 26, 2021**Accepted:** August 18, 2021**Article in press:** August 18, 2021**Published online:** September 18, 2021**P-Reviewer:** Liu J**S-Editor:** Yan JP**L-Editor:** Filipodia**P-Editor:** Zhang YL

Core Tip: Timely diagnosis of biliary atresia (BA) is critical to optimizing the outcomes of Kasai procedure (KP), which should be performed as early as possible. Children with a delayed diagnosis of BA at high risk of early KP failure or those presenting with clear evidence of decompensated cirrhosis should be considered for primary liver transplantation (LT). Early KP failure requiring salvage LT within the first 2-3 years of life occurs in nearly half of all children with BA but even those with a successful KP need life-long monitoring for progression of liver disease that may require salvage LT.

Citation: Kakos CD, Ziogas IA, Alexopoulos SP, Tsoulfas G. Management of biliary atresia: To transplant or not to transplant. *World J Transplant* 2021; 11(9): 400-409

URL: <https://www.wjgnet.com/2220-3230/full/v11/i9/400.htm>

DOI: <https://dx.doi.org/10.5500/wjt.v11.i9.400>

INTRODUCTION

Biliary atresia (BA) represents the most common indication for pediatric liver transplantation (LT) worldwide, accounting for half of LTs in children and one-tenth of all LTs[1]. Kasai was the first who performed successful drainage of the bile into the intestine after resecting the obliterated extrahepatic portion of the biliary tree[2].

Although the Kasai procedure (KP) is considered to be the first-line treatment, the progressive liver injury seen in most patients with BA results in a 5-year post-procedural native liver survival (NLS) between 38%-40% even at experienced centers [3-5]. As a result, most patients will need a salvage LT (sLT) at some point during their lifetime. The overall low lifetime NLS creates an important dilemma for pediatric LT experts: Should LT be considered primary therapy in infants with BA and a high likelihood of KP failure, or should it be utilized as a salvage therapy? This review aims to highlight the key concepts around this question and to provide an update regarding the management and outcomes of KP and LT for BA.

PATHOPHYSIOLOGY

The pathogenesis of BA is not fully understood but appears to be multifactorial. Approximately 15% of patients with BA have associated congenital malformations, such as abdominal and thoracic heterotaxia, polysplenia, asplenia, and intestinal malrotation[6]. Viruses, such as cytomegalovirus, herpes virus, Epstein-Barr virus, and reovirus, may also contribute to a certain extent in the pathogenesis of BA. Additional factors which may contribute are neonatal immune dysregulation and environmental toxins[7,8]. While radiologic studies, such as hepatobiliary iminodiacetic acid scan, may suggest the diagnosis, failure to visualize the biliary tree during intra-operative cholangiography remains the gold standard for diagnosing BA. Characteristic findings on liver biopsy include edematous fibroplasia with bile ductular proliferation and bile plugs[9].

FACTORS ASSOCIATED WITH NATIVE LIVER SURVIVAL AFTER KASAI

Before the Kasai's report[2], BA was a fatal disease. Unfortunately, KP does not prevent progressive hepatic injury, which gradually leads to cirrhosis and end-stage liver disease (ESLD) in most patients. Numerous studies have attempted to identify the factors predictive of NLS, however, the majority are from single centers and retrospective in nature[1,3,10-15]. Additionally, many of them are limited to univariate analysis, and thus careful interpretation of these results is warranted (Table 1).

The main histologic characteristics of BA are increased cholestasis, marked fibrosis, and ductular proliferation, while the mechanisms behind fibrogenesis are still under investigation. There seemed to be a clear association between the degree of preoperative fibrosis in the liver biopsy and poor NLS[10,16-18]. Another important histologic finding is intrahepatic duct size, as ducts less than 200 µm represent a risk factor for NLS[19]. However, a large prospective study from 16 centers in North

Table 1 Factors reported to be associated with native liver survival

Before the Kasai procedure	After the Kasai procedure	Other
Liver fibrosis	Jaundice clearance	Specialized institution
Ductal size	Cholangitis	
Biliary atresia type according to Ohi classification	Total bilirubin	
Portal hypertension	Serum creatinine	
Biliary atresia splenic malformation syndrome	Portal hypertension	
Age at the time of Kasai procedure	Serum albumin	
	Corticosteroids	
	Antibiotics	

America found no association between liver fibrosis severity, measured histologically by the 6 grade Ishak score, and NLS. Instead, gross appearance of the liver at the time of surgery was predictive of poor NLS[20]. Regarding BA types based on the Ohi classification, patients with Ohi type 2 and 3 BA appear to have worse outcomes than Ohi type 1[20].

Duché *et al*[21] showed that elevated portal pressure, polysplenia syndrome, and complete atresia of the extrahepatic biliary remnant were independently associated with worse NLS, as there were lower chances of successful postoperative jaundice clearance. The latter is considered extremely important for a proper KP[22,23]. Superina *et al*[20] have also shown the effect of early jaundice clearance on improved NLS. Similar to Duché *et al*[21], Superina *et al*[20] reported the hazardous effect of BA splenic malformation syndrome (BASM) on NLS, which surprisingly was not associated with jaundice clearance after KP. The presence of splenic anomalies as an indicator of poor prognosis has also been documented in other studies[3,24]. The embryological aspects of this malformation have been studied by Davenport *et al*[25] and Karrer *et al*[26], yet the pathogenesis is still unclear. Notably, a study from Sendai, Japan reported similar survival between patients with isolated BA *vs* BA plus BASM and no associated cardiac defects[27]. Sasaki *et al*[28] demonstrated that presence of symptomatic portal hypertension (gastro-esophageal varices requiring treatment), but not hypersplenism nor cholangitis, was found to be a significant risk factor in multivariate logistic regression analysis for NLS.

Age at the time of KP plays a vital role in NLS[5]. There is a general consensus among pediatric surgeons that the sooner the diagnosis and KP is performed, the better the outcome. Several cutoffs between 7-10 wk after birth have been proposed in the literature[3,10,20,29]. It is noteworthy that the age at KP in the United States has not decreased significantly over time[30].

Expertise in KP offered in high-volume centers and centralization of care for patients with BA has been thought of playing a key role in improving outcomes. Although excellent results can be obtained even in centers with relatively little experience[31], this theory seems to have a strong basis[3]. The so-called “center effect” reflects the experience of the teams at individual centers, and in certain countries in Europe (*e.g.*, United Kingdom, Finland) centralization of care to supraregional centers has been effective in optimizing outcomes nationwide[32,33].

Regarding postoperative factors, recurrent cholangitis episodes have been associated with KP failure in multiple studies[18,34-36]. More specifically, Wildhaber *et al*[18] demonstrated an approximately double risk for patients with bridging fibrosis and postoperative cholangitis compared to those with cholangitis only, showing the impact of this underlying condition. Moreover, Wu *et al*[35] noted that patients with BA and inadequate bile drainage had more cholangitis episodes than those with adequate bile drainage, while the occurrence of cholangitis was associated with decreased NLS in both groups of patients. In contrast, a single-center study from California showed no association between cholangitis episodes and the need for LT [19], and the same was reported in a more recent study from Japan[28]. It is well-established that BA is an obstructive intra- and extrahepatic cholangiopathy, while KP can only solve the extrahepatic part of the problem. Therefore, for optimal outcomes, KP can be combined with regimes dealing with intrahepatic obstruction, inflammation, and bile infection[37]. Specifically, ursodeoxycholic acid has been utilized often for this purpose due to its immunomodulatory and cytoprotective effects[38].

Jain *et al*[39] described several parameters associated with an increased risk for requiring LT after 16 years of age. They reported that among BA patients achieving NLS until the age of 16 years, only serum total bilirubin and creatinine were associated with higher risk of requiring LT. A retrospective study from Australia and Canada showed that a serum albumin level below 35 g/L was a poor prognostic indicator in infants with BA who were no longer jaundiced at 3 mo after KP[40].

Corticosteroids are well-known modulators of BA inflammation as they reduce the production of inflammatory cytokines (tumor necrosis factor- α , interleukin-1, interleukin-8), prostaglandins, and nitric oxide[41]. They also seem to have other choleretic effects, which have not been studied as extensively. Results from early studies on the use of corticosteroids showed a benefit in survival in BA patients[42-45]. A randomized controlled trial from 2007 showed that corticosteroids had a benefit on the rate of reduction of bilirubin early postoperatively, yet they did not reduce the need for LT[46]. The more recent START randomized clinical trial compared 70 children receiving intravenous methylprednisolone (4 mg/kg/d for 2 wk) and oral prednisolone (2 mg/kg/d for 2 wk) followed by a tapering protocol for 9 wk with 70 children receiving placebo initiated within 3 d of KP[47]. The study showed that high-dose steroids after KP did not significantly improve bile drainage at 6 mo, although a small clinical benefit could not be excluded[47]. However, treatment with steroids was associated with earlier onset of serious adverse events[47]. A meta-analysis published in 2015 showed no significant difference in jaundice clearance for patients who received steroids overall; nonetheless, sensitivity analysis excluding studies on the use of high- or low-dose steroids and including only studies on the use of moderate-high dose steroids (prednisolone 4-5 mg/kg/d) showed a higher jaundice clearance rate at 6 mo post-KP[48]. Prednisolone is the most frequently prescribed steroid in most studies, but dexamethasone and hydrocortisone have also been described in the literature[49]. The possible side effects of long-term steroid use should not be neglected.

There is limited knowledge about potential benefit of post-KP use of antibiotics. The commonest intravenous regimen in a survey of European practice is a combination of piperacillin-tazobactam and gentamicin[50]. A randomized clinical trial by Bu *et al*[51] demonstrated a positive impact of post-KP use of trimethoprim-sulfamethoxazole or oral neomycin, while a recent systematic review of four articles by Dechaurun *et al*[52] presented ambiguous results with three studies suggesting the presence of a potential benefit in using antibiotics. The need for high-quality evidence in the form of prospective studies in this field is evident.

LT OUTCOMES

The majority of patients with BA will progress to ESLD requiring evaluation for LT at some point in their life. Since Kasai's first description[2], there have not been significant changes to the technique of KP, and long-term NLS has not significantly improved. Unfortunately, even patients who manage to survive more than 20 years after KP have histological, clinical, or ultrasonographic evidence of significant chronic liver disease[53,54]. Portal hypertension is also commonly observed in BA patients at some point after KP[53,54].

sLT is considered when patients who had undergone KP develop ESLD. A retrospective study from the United States reported a higher incidence of cholangitis, sepsis, and bacteremia in the baseline characteristics of patients who underwent sLT, compared with those who underwent only KP[55]. The authors also compared the outcomes between primary liver transplantation (pLT) and KP, regardless of whether patients eventually required sLT. Early survival was higher in the KP group, but long-term survival was significantly better in pLT group (5-year survival 88% for KP *vs* 94% for pLT)[55]. sLT was also associated with an increased risk of death compared to pLT, which may be attributed to the technical difficulties of sLT in the setting of previous KP and hilar dissection. Recipients of sLT for BA have been reported to have a higher incidence of infectious and vascular complications and intestinal perforation compared to pLT recipients, likely due to previous surgical interventions[56-58]. The incidence of pLT for BA varies from 10%-11% in Canada, Switzerland, and Germany to 3%-4% in the Netherlands, United Kingdom, and France and to 0.1% in Japan[59], so the decision regarding the management of BA may vary among different healthcare systems.

Nevertheless, there are studies reporting equivalent LT outcomes between patients with and without a previous KP. The findings of equivalent post-LT survival

regardless of prior KP support the recommendation for a staged approach for the treatment of BA, starting with KP and progressing to LT only when necessary[60-63]. It is argued that in this way, KP delays the need for LT and allows not only for the improvement of the child's nutritional status but also for their size to increase and to increase the potential size-matched organ donor pool. A multicenter study from 39 centers in the USA and Canada failed to demonstrate an effect of prior KP on LT outcome[64]. Cowles *et al*[65] reported on 71 children who underwent LT for BA, 61 of whom had previously undergone KP, and they observed no clear difference in the outcomes between the two groups. A 2016 meta-analysis reported no difference in 1- and 5-year patient and graft survival between patients who underwent KP and those who did not, yet patients who had undergone KP prior to LT had an increased risk of postoperative infection[66].

Another interesting aspect is the comparison of post-transplant after KP outcomes between children and adults. Kyoden *et al*[67] found no significant differences in survival with a 5-year patient survival of 90% in both age groups, yet a large retrospective study from Japan demonstrated a clear survival benefit in the pediatric population (5-year patient survival 86.7% in children *vs* 69.7% in adults)[68]. In both studies the patients received a living donor graft[67,68]. A more recent single-center study from King's College Hospital also showed superior patient survival after deceased donor LT for BA patients listed as children ($n = 22$) compared to those listed as adults ($n = 14$), yet the results did not reach statistical significance because of the limited study sample[69].

The type of liver graft may also be a major prognostic factor. Living donation has expanded the donor pool and also provided recipients with organs which appear to be of better quality than the deceased ones[70]. Multiple studies agree that living donor grafts have superior outcomes in patients with BA[60], but there are also reports challenging this theory[61,63,64]. A study from 1996 had suggested that LT using reduced-size grafts (only part of the donor liver is used for the graft, and the remaining resected liver is discarded) may not be the best option for BA due to inferior outcomes[62]. However, since reduced-size grafts were mostly utilized in emergency situations, the authors stated that after censoring these, they found no significant difference in patient survival between elective reduced-size and whole liver grafts[62]. A more recent study using national registry data showed that the effect of donor allograft is related to recipient weight for children with BA[71]. Specifically, for children ≤ 7 kg, reduced size grafts and living donor grafts had decreased risk of graft failure compared to whole grafts, for children 7-14 kg living donor grafts had decreased risk of graft failure compared to both reduced size and whole grafts, while for children > 14 kg there was no difference in graft failure by allograft type[71].

There are several studies that tried to identify predictors of a successful LT in children with BA (Table 2). Fouquet *et al*[61] demonstrated that BASM, intraoperative complications (hemorrhage, intestinal injury, vascular thrombosis), and hospitalization in the intensive care unit were associated with an increased risk of death. Uttersen *et al* [64] reported that infant recipient (≤ 11 mo), use of cyclosporine *vs* tacrolimus, growth deficit, and re-transplantation were associated with post-LT mortality. Living donation, technological refinements, increased surgical experience, and advances in anesthesia and in immunosuppression will play a key role in improving post-LT outcomes.

TRANSITION OF CARE

So, the main question remains: To transplant or not to transplant? For many decades it was considered that KP must be the first choice for patients with BA, serving as a bridging therapy to delay or even avoid the need for LT. Today it is well-established that most BA patients will eventually require LT at some point in their lifetime, yet the demand for donor livers continuously exceeds the supply. LT has matured to the stage where in most centers excellent long-term survival can be achieved despite the technical challenges of sLT following KP. However, the excellent long-term outcomes of pLT suggest this is also a reasonable alternative treatment option for certain patients [59]. pLT is now being more frequently considered for children with BA at a very high likelihood of early failure of KP, challenging the traditional treatment paradigm.

For children who have undergone KP, the best next step is to ensure adequate follow-up and appropriate transition of care from childhood to adulthood so as to continuously monitor for manifestations of ESLD and refer for LT when needed. Progressive jaundice, recurrent bacterial cholangitis, portopulmonary hypertension,

Table 2 Factors associated with liver transplant outcomes

Patient characteristics	Surgical characteristics
Age at the time of liver transplant	Previous Kasai procedure
Biliary atresia splenic malformation	Intraoperative complications (hemorrhage, intestinal injury, vascular thrombosis)
Growth deficit	Allograft type
Hospitalization in the intensive care unit	Re-transplantation
Type of immunosuppression	

and hepatopulmonary syndrome warrant evaluation for LT. Implementation of objective scoring systems, including Model for End-stage Liver Disease and Pediatric End-stage Liver Disease score systems, have decreased the pediatric waitlist mortality and increased the number of patients receiving a deceased donor liver graft[55]. However, several manifestations of ESLD are not adequately reflected in these scoring systems, and thus many children with BA eventually require exception points to undergo LT[72]. Mortality risk has been shifted gradually to the pre-transplant period, and peri-transplant risks are mainly related to patient's condition[73]. Receiving an LT at a young age allows for greater use of left lateral segment graft from a living donor without affecting the deceased donor pool. From another point of view, KP is far more cost-effective compared to pLT[74].

Timely diagnosis of BA is critical to optimizing the outcomes of KP, which should be performed as early as possible. Children with a delayed diagnosis of BA at high risk of early KP failure or those presenting with clear evidence of decompensated cirrhosis should be considered for pLT. Early KP failure requiring sLT within the first 2-3 years of life occurs in nearly half of all children with BA, but even those with a successful KP need life-long monitoring for progression of liver disease that may require sLT.

CONCLUSION

In conclusion, cooperation between pediatric and adult hepatologists, pediatric surgeons, and transplant surgeons is necessary for the management of BA patients. Close and long-term follow-up is required to monitor for manifestations of ESLD that may warrant evaluation for LT, as well as to improve quality of life along with survival outcomes. More prospective multicenter studies are needed to demonstrate a clear conclusion about the proper management of BA.

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