Check for updates

Pediatric Transplantation

WILEY

ORIGINAL ARTICLE

Primary Liver Transplantation for Biliary Atresia: Another Piece of the Puzzle

Jean de Ville de Goyet[1,2] | Giovanni Boroni[3] | Daniela Liccardo[4] | Daniele Alberti[3] | Marco Spada[5] | Valeria Casotti[6] | Raffaele Iorio[7] | Pier Luigi Calvo[8] | Nicolo Rizzitello[9] | Giusy Ranucci[1,2] | for the Italian Society of Paediatric Gastroenterology, Hepatology, and Nutrition (SIGENP) AVB Network Working Group

[1]Paediatric Liver Unit, ISMETT, Palermo, Italy | [2]UPMC Italy, Palermo, Italy | [3]SCDU Chirurgia Pediatrica, ASST Spedali Civili di Brescia—Ospedale Dei Bambini, Brescia, Italy | [4]Division of Hepatology and Metabolic Diseases, Liver Transplant Unit, Bambino Gesù Children's Hospital, IRCCS, Rome, Italy | [5]Division of Hepatobiliopancreatic Surgery, Liver and Kidney Transplantation, Bambino Gesù Children's Hospital, IRCCS, Rome, Italy | [6]Paediatric Hepatology, Gastroenterology and Liver Transplant Unit, Ospedale Papa Giovanni XIII, Bergamo, Italy | [7]UOS Epatologia Pediatrica, Università di Napoli Federico II, Napoli, Italy | [8]Gastroenterologia Pediatrica. Ospedale Infantile Regina Margherita, Azienda Ospedaliero-Universitaria Città Della Salute e Della Scienza di Torino, Turin, Italy | [9]Statistics and Data Management Services, ISMETT, Palermo, Italy

Correspondence: Jean de Ville de Goyet (jdeville@ismett.edu)

Received: 28 May 2026 | **Revised:** 29 August 2026 | **Accepted:** 5 September 2026

Keywords: biliary atresia | national registry | outcome | primary liver transplantation

ABSTRACT

Background: Primary liver transplantation (PLT) remains a controversial alternative strategy for children with biliary atresia (BA). This national study of BA management in Italy over a 10-year period aimed to better define the indications, limitations, and outcomes of PLT.

Methods: This was a retrospective, multicenter, observational study based on the Italian BA Registry, including 312 infants with BA managed in Italy between January 2012 and December 2021. All children who underwent PLT were selected for analysis. Children who underwent liver transplantation (LT) after Kasai portoenterostomy (KPE) failure were used as a comparison group.

Results: Primary liver transplantation was performed in 37 children with BA and established advanced liver disease; 181 other BA patients who underwent LT were selected for comparison. Minimum follow-up was 3 years for all patients. PLT infants were significantly older at diagnosis (13 were diagnosed after 120 days of life). They had a significantly worse clinical condition both at diagnosis and at listing for transplantation. No child died before LT in the PLT group, whereas 5 patients died before LT in the early rescue-LT group. Post-transplant outcomes were comparable across groups.

Conclusions: Kasai portoenterostomy should remain the recommended first-line treatment for most infants with BA, but its benefit is limited to only a proportion of patients. Some patients may be processed rapidly to LT because they present too late or with advanced disease: these patients have excellent outcomes as well.

1 | Introduction

Kasai portoenterostomy is the standard initial treatment for BA and remains the only procedure aimed at restoring bile flow and preserving the native liver. Without KPE, untreated BA progresses inexorably to biliary cirrhosis and end-stage liver failure [1–4]. However, KPE is not an absolute cure: it fails in at least one-third of patients during the first year after surgery, and

Abbreviations: BA, Biliary Atresia; IQR, Interquartile range; KPE, Kasai Portoenterostomy; LT, Liver transplantation; PLT, Primary liver transplantation. SIGENP representative (https://www.avb-sigenp.org/site/home (see also Table S1)).

© 2026 Wiley Periodicals LLC.

All rights reserved, including rights for text and data mining and training of artificial intelligence technologies or similar technologies.

long-term native liver survival decreases progressively, reaching about 25%–30% at 10 years in many series.

In this context, PLT has periodically been proposed as an alternative strategy, with the rationale of offering a treatment in a single step and avoiding deterioration associated with progressive cirrhotic liver disease. This approach remains controversial because KPE still offers native liver survival to a substantial proportion of infants, and previous KPE does not appear to compromise subsequent LT outcomes in experienced centers.

The Italian Biliary Atresia Network was established in 2018 under the auspices of the Italian Society of Pediatric Gastroenterology, Hepatology, and Nutrition and created a centralized platform collecting data on Italian BA patients from 2012 onward (https://www.avb-sigenp.org/site/home) [5]. The present report describes the retrospective analysis of this registry over a 10-year period, focusing specifically on the role of PLT in the management of BA.

2 | Patients and Methods

2.1 | Study Design and Ethics

This was a retrospective, observational, multicenter national study based on the Italian Biliary Atresia Registry: the registry had collected retrospectively the information about all children diagnosed with BA in Italy between January 2012 and December 2021, who had been managed and cured in the 13 participating centers of the Network; in Italy, the Health Ministery does not designate national centers, as health care is regionalized: BA management, as any other care, follows a decentralized care model. Details of participating centers were reported in Supporting Information in the draft manuscript.

This analysis focused on children managed with PLT and was conducted in parallel with a larger study of BA patients undergoing KPE during the same period. For this study, only 7/13 network centers contributed as the other centers didn't manage PLT cases; these 7 centers were the largest centers of all, having enrolled altogether 81% of the patients of the registry. Of note, all consecutive PLT performed in the study period have been identified, as the former 7 centers included all Italian pediatric transplant centers.

The study was approved by the SIGENP scientific board (NP3854), the National Italian Ethics Committee, and the local ethics committee of each participating center. Parents or legal representatives provided informed consent for inclusion in the registry and participation in the study. The protocol conformed to recognized ethical standards and to the ethical principles of the Declaration of Helsinki. Informed consent was obtained prospectively, covering the retrospective use of registry data for clinical research purposes and scientific publication.

2.2 | Patient Selection

All patients with confirmed BA who were registered between January 2012 and December 2021 and were proposed for PLT were included as Group 1. For comparison, all infants in the registry who underwent LT after KPE were identified as "failed KPE" and divided into 2 groups according to timing of LT. Group 2 included infants who underwent early LT for failed KPE, defined as LT before 485 days of age, a threshold selected to best match the age distribution of LT in the PLT group. Group 3 included registry patients who underwent late LT after 485 days of age. The cut-off of 485 days was decided on the basis that all (but one outlier [6]) PLT were performed in infants aged up to 485 days.

To analyze the effect of decentralized care as it is in Italy (BA diagnosis and KPE in non-designated centers), the native liver survival (days) and the age at listing (days) were compared between two groups as follows: group A consisted of all children who had their KPE and their transplant in the same (transplant) center, and group B was formed of all other patients (who had their management and/or their operations in different centers).

Collected variables included demographic data, clinical information before and around surgery, anatomical and histological findings, and follow-up information.

2.3 | Statistical Analysis

For descriptive and exploratory analyses, categorical variables were summarized as counts and percentages, whereas continuous variables were described as mean and standard deviation when normally distributed, or as median and interquartile range (IQR) otherwise. Group comparisons were performed with analysis of variance and ANOVA/Tukey testing for normally distributed continuous variables, with Kruskal–Wallis test for non-normally distributed variables, and with the chi-square test or Fisher exact test for categorical variables. Statistical analyses were performed using SAS version 9.4. All tests were two-sided, and a p value ≤ 0.05 was considered statistically significant.

3 | Results

Among 312 BA patients registered in the Italian BA Registry, 37 infants (11.9%) underwent PLT (Group 1). A further 181 transplanted patients were selected for comparison: 127 underwent KPE followed by early rescue-LT before 485 days of age (Group 2), and 54 underwent late LT after 485 days of age (Group 3). Minimum follow-up for all patients was 3 years (Figures 1 and 2).

In the PLT group, 11 infants underwent exploratory laparotomy without KPE. KPE was not performed because of advanced cirrhosis documented (macroscopically or by liver biopsy in all 11), with clinical ascites in 8, or portal vein agenesis with cavernomatous transformation in 1. In the latter group, exploration had been proposed after Ultrasonography had not anticipated clearly the presence of a contraindication for KPE—decision that was made in fact extemporaneously by the surgeon. Two of the 37 PLT patients had other congenital malformations and had previously undergone abdominal surgery in the first days of life (1 jejunal and 1 ileal atresia) with the liver disease (then at a very early stage) being not recognized.

13993046, 2026, 10, Downloaded from https://onlinelibrary.wiley.com/doi/10.1111/petr.70458 by Universita Di Brescia, Wiley Online Library on [06/11/2026]. See the Terms and Conditions (https://onlinelibrary.wiley.com/terms-and-conditions) on Wiley Online Library for rules of use; OA articles are governed by the applicable Creative Commons License

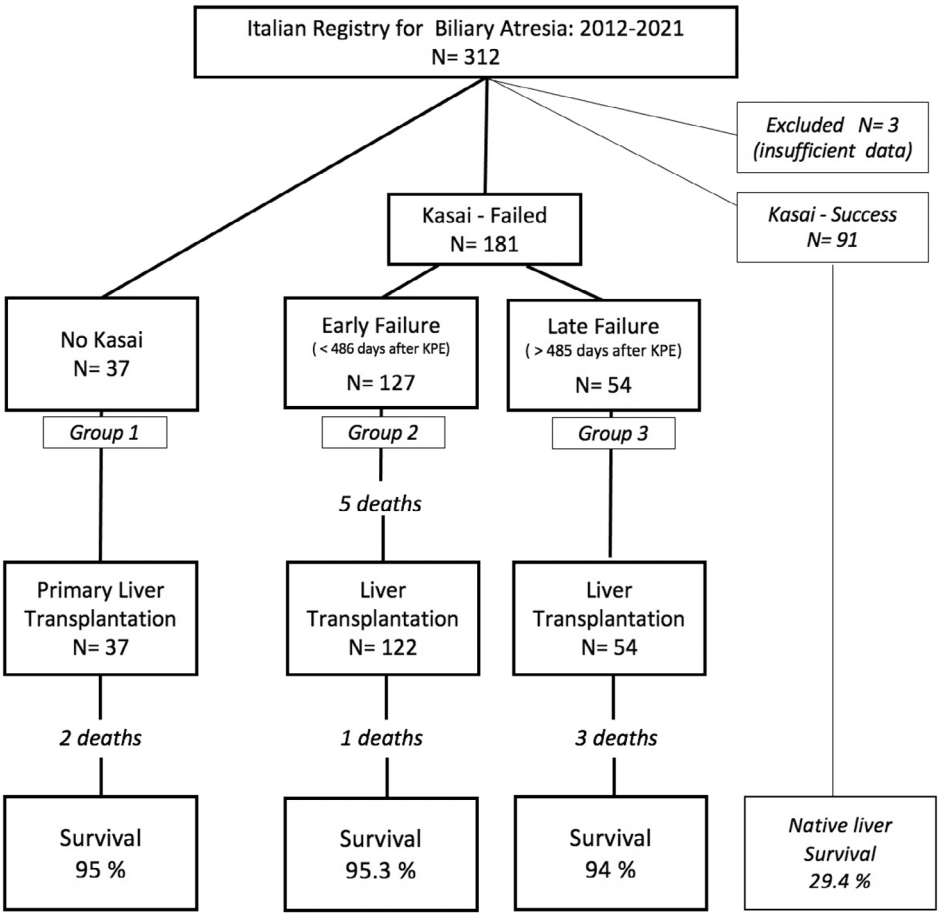


FIGURE 1 | Flow diagram of the cohort, subgroups and outcomes.

PLT infants were significantly older at diagnosis, with a mean age of 108 days, and 13 were diagnosed after 120 days of life (Table 1). Late diagnosis was associated with a worse clinical condition at initial assessment, including more advanced hepatopathy and significant differences in direct bilirubin, ALT, GGT, and INR compared with the other groups (Table 1). PLT infants were younger and smaller at the time of listing for LT compared to those in the other groups; surprisingly, they also had significantly higher PELD scores (Table 1 and Figure 3). Despite the early decision to proceed to PLT (decision made at diagnosis), 27 of these 37 infants had developed refractory ascites by the time of listing, and 18 had severe coagulopathy (Table 1).

Waiting time was similar in Groups 1 and 2 (Table 1). No death occurred before LT in the PLT group, whereas 5 patients died before transplantation in Group 2. Five children in Group 2 died before transplantation. One child died at age 18.5 months during an acute episode of hemorrhage: he had been on the list for 4.5 months. Four infants died before the age of one year, in relation to prematurity with chromosomal abnormalities ($N=1$) or multiple anomaly syndrome ($N=1$), acute hemorrhage ($N=1$) and RSV infection ($N=1$).

Two PLT patients died after LT (95% survival rate), a rate comparable to that observed in the other transplanted groups (Figures 1 and 4).

To analyze the possible effect of decentralization, the age at listing (days) and the native liver survival (days) were compared between group A (children who had KPE and transplant in the same center—63% of cases) and group B (all others—37% of patients). The median (IQR1-3) age at listing was 234 (176–343) days and 254 (169–487) days, respectively in group A and B ($p=0.74$). The median (IQR1-3) survival time of the native liver was 210 (154–413) days and 281 (157–569) days, respectively in group A and B ($p=0.75$).

4 | Discussion

In this national registry-based study, PLT accounted for about 11% of BA management in Italy between 2012 and 2021. This rate lies within the broad range reported worldwide. Reported incidences of PLT in the world vary between 0.5%–16% [7–16](only one outlier Brazilian center reporting rates as high as 40% [16]). Two series (Japanese registry and England/Wales program) report rates as low as 0.5% and 2.9%, respectively—likely reflecting a combination of earlier diagnosis, an aggressive KPE policy even in late-presenting infants, and limited deceased-donor organ availability [7, 17, 18]. In this cohort, PLT was mainly reserved for infants who presented late with advanced liver disease, often with associated malformations. Splenic Malformation syndrome (BASM) was overrepresented in the PLT group (22% vs. 9%, $p=0.030$, Table 1).

13993046, 2026, 10, Downloaded from https://onlinelibrary.wiley.com/doi/10.1111/petr.70458 by Universita Di Brescia, Wiley Online Library on [06/11/2026]. See the Terms and Conditions (https://onlinelibrary.wiley.com/terms-and-conditions) on Wiley Online Library for rules of use; OA articles are governed by the applicable Creative Commons License

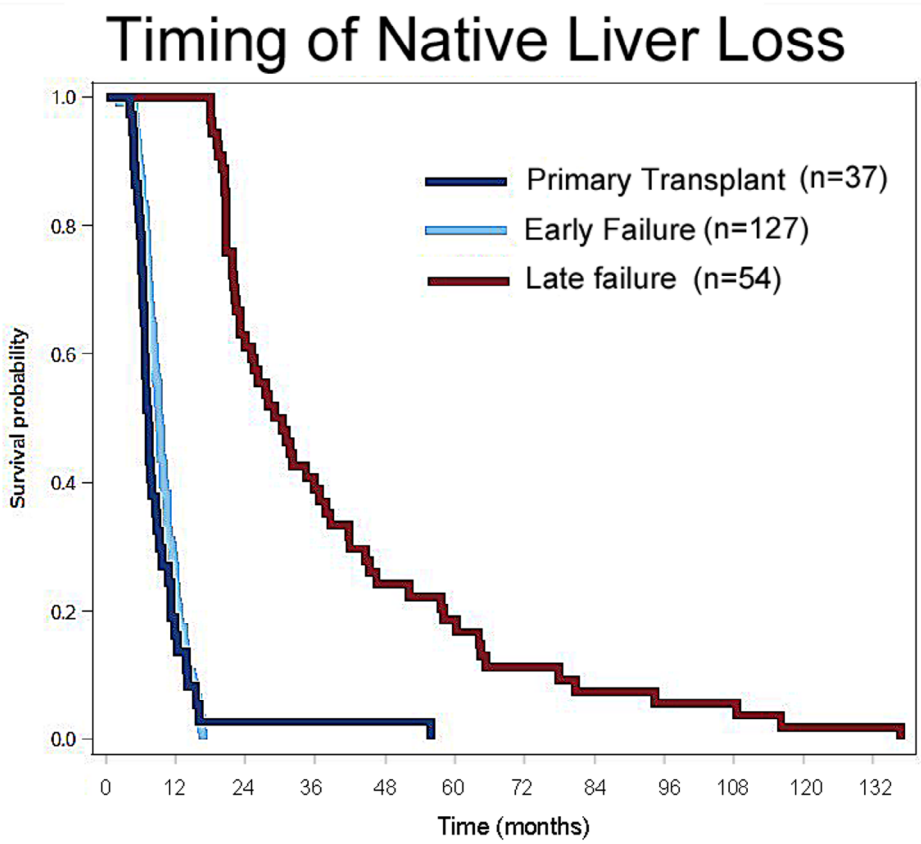


FIGURE 2 | Timing of Native liver loss in a cohort of 218 Biliary Atresia children who underwent liver transplantation (Italy 2012–2022), according to clinical indication: Primary liver transplantation (no Kasai portoenterostomy), early failure of Kasai (transplant within 485 days of age), or late failure after Kasai (> 485 days of age).

In our cohort, at diagnosis, PLT candidates had a significantly worse clinical profile than infants undergoing KPE followed by rescue LT, with more severe cholestasis, coagulation abnormalities, and liver injury. This difference persisted and widened over time, as PLT infants had significantly higher PELD scores at listing and more frequent refractory ascites and severe coagulopathy. Thus, PLT in this series was not a strategy chosen for convenience or preference, but rather one reserved for a particularly sick subgroup of patients.

Despite this severe baseline profile, no PLT infant died on the waiting list, whereas 5 deaths occurred among infants listed after early KPE failure; although the latter is not significantly different, it doesn't seem to incentivize against PLT per principle—mostly indicating the need for further studies. This observation suggests that early KPE failure may define a subgroup at particularly high risk of pretransplant deterioration and death, even when their condition at diagnosis appears less severe than that of infants selected for PLT. At the same time, it highlights a paradox in the PLT pathway: although the decision to forgo KPE had already been made at diagnosis (median 108 days), listing for LT was only completed at a median of 169 days (an interval of roughly 2 months), during which clinical condition continued to worsen (Table 1 and Figure 3).

These results are in line with recent literature showing that LT outcomes for BA are now excellent whether transplantation is primary or follows KPE (Figure 4) [19–25]. Contemporary series report patient survival after PLT above 90%, with similarly favorable outcomes after sequential LT [22–24], while earlier reports showed wider variability and higher pretransplant mortality. These data also support the established concept that KPE does not compromise subsequent LT outcomes in experienced centers, reinforcing its place as first-line treatment for most BA infants.

Nevertheless, the present data raise the important question of whether all BA infants should continue to follow the same KPE-first pathway. A substantial subset of children derives little long-term benefit from KPE and ultimately proceeds to urgent LT in poor condition after early failure. For these infants, earlier identification of poor KPE candidates could allow more appropriate transplant-oriented strategies. Although literature brings increasing information about prognostic markers for a successful KPE, the latter studies analyze either histology at KPE or blood parameters after KPE; until now there is little or nothing that has been proposed for predicting -with enough sensitivity and specificity- a successful KPE ahead of the operation. As far as the research is today, there is no test that would allow selecting the right candidates for successful KPE, while proposing others to PLT for high likelihood of KPE failure. Further research is on the way and is necessary before one can develop a new model for managing BA more selectively.

From a pediatric transplant perspective, several practical implications emerge. Earlier listing should be considered once the

13993046, 2026, 10, Downloaded from https://onlinelibrary.wiley.com/doi/10.1111/petr.70458 by Universita Di Brescia, Wiley Online Library on [06/11/2026]. See the Terms and Conditions (https://onlinelibrary.wiley.com/terms-and-conditions) on Wiley Online Library for rules of use; OA articles are governed by the applicable Creative Commons License

TABLE 1 | Clinical characteristics of biliary atresia patients proposed for primary liver transplantation (No Kasai), or for sequential Kasai Porto-enterostomy (KPE) and rescue liver transplantation – either for early or late failure of KPE.

	1 No Kasai ($n=37$)	2 KPE early failure ($n=127$)	3 KPE and Late LT ($n=54$)	p-value (1 vs. 2)	p-value (1 vs. 3)	p-value (2 vs. 3)
AT diagnosis						
Gender (F)	16 (43%)	78 (61%)	34 (63%)	0.049	NS	NS
Gestational age (weeks)	38.2 (2.2)	38.6 (1.7)	38.5 (2.1)		NS	
Age at diagnosis (days)	108 (37)	57 (24)	53.5 (25,8)	< 0.0001	< 0.0001	NS
Total bilirubin (mg/dL)	12.5 (6.7)	9.5 (3.1)	10.5 (3.9)	0.0004	NS	NS
Conjugated bilirubin (mg/dL)	9.3 (4.2)	6.8 (2.3)	7.7 (3.1)	< 0.0001	< 0.0001	NS
AST (IU/dL)	353 (252–505)	179 (144–301)	226 (125–327)	0.0007	0.0007	NS
ALT (IU/dL)	213 (152–314)	139 (88–223)	167 (97–206)		NS	
GGT (IU/dL)	353 (226–680)	515 (292–850)	626 (333–1240)	0.0073	0.0073	0.007
INR (ratio)	1.2 (1.0–1.4)	1.0 (1.0–1.1)	1.0 (1.0–1.1)	0.0269	0.0269	NS
Biliary Atresia Splenic malformation syndrome (n)	8 (22%)	11 (9.1%)	9 (17%)	0.030	NS	NS
Age at KPE (days)	na	72 (56–83)	65 (47–82)		na	NS
Steroids after KPE	na	76 (60%)	40/50 (80%)		na	0.01
At listing for transplantation						
Age at listing (days)	169 (147–205)	219 (164–274)	681 (517–1222)	< 0.0001	< 0.0001	< 0.0001
Weight at listed for LT	6.4 (5.9–6.9)	6.9 (6.0–7.8)	12,1 (9.9–14.1)	< 0.0001	NS	< 0.0001
PELD score	21.9 (7.1)	17.9 (6.8)	9.7 (7.0)	< 0.0001	< 0.0001	< 0.0001
INR (ratio) at listing	1.7 (1.4–2.0)	1.4 (1.2–1.8)	1.2 (1.1–1.4)	< 0.0001	NS	< 0.0001
Total Bilirubin (mg/dL)	19.8 (9.7)	17.1 (9.8)	9.7 (7.6)	NS	0.0027	0.020
Conjugated bilirubin (mg/dL)	16.2 (8.2)	13.1 (9.3)	8.2 (6.8)	NS	0.017	NS
Severe Ascites[a]	27 (73%)	75 (67%)	13 (28%)	NS	< 0.0001	< 0.0001
Diuretic therapy	31 (89%)	88 (79%)	22 (47%)	NS	< 0.0001	< 0.0001
Severe coagulopathy[b]	18 (49%)	37 (30.6%)	4 (7%)	0.012	< 0.0001	0.0014

(Continues)

13993046, 2026, 10, Downloaded from https://onlinelibrary.wiley.com/doi/10.1111/petr.70458 by Universita Di Brescia, Wiley Online Library on [06/11/2026]. See the Terms and Conditions (https://onlinelibrary.wiley.com/terms-and-conditions) on Wiley Online Library for rules of use; OA articles are governed by the applicable Creative Commons License

TABLE 1 | (Continued)

	1 No Kasai ($n=37$)	2 KPE early failure ($n=127$)	3 KPE and Late LT ($n=54$)	*p*-value (1 vs. 2)	*p*-value (1 vs. 3)	*p*-value (2 vs. 3)
Cholangitis episode	—	29 (23%)	19 (35%)	na		NS
Waiting time (days)	44 (22–99)	41 (19–84)	86 (27–298)	NS	< 0.0001	< 0.0001
At transplantation						
Age at LT (days)	217 (181–322)	274 (211–355)	898 (6554–1410)	NS	< 0.0001	< 0.0001
Death before transplant	—	5 (3.9%)	—		NS	
Death after transplant	2 (5%)	1 (0.8%)	3 (6%)		NS	
Patient survival	35 (95%)	121 (95.3%)	51 (94%)		NS	

Note: ANOVA and Tukey Test, or Chi-Square (C2) comparisons. Values are expressed as Mean (SD), or as Median (IQR1-IQR3), or as *n*(%).
Abbreviation: LT, liver transplantation.
[a]Severe Ascites defined as presence of ascites despite diuretic treatment.
[b]Severe coagulopathy defined as INR ratio > 1.7.

FIGURE 3 | Age and PELD score at registration on the list for Liver Transplantation in two groups of infants with Biliary Atresia. Patients proposed to LT after failed Kasai portoenterostomy (sequential LT) (blue) and patients having primary transplantation (Primary LT) (orange).

decision for PLT has been made rather than waiting for weight gain or further stabilization outside the waiting list. Intensive nutritional and medical support while awaiting LT is essential, and broader use of split grafts, living donor transplantation, ABO-incompatible transplantation in selected infants, and advanced graft-reduction techniques may further improve access to timely LT in this vulnerable population. These approaches are particularly relevant for high-volume pediatric transplant centers.

Future work should focus on defining robust preoperative predictors of KPE failure, integrating clinical, biochemical, imaging, and composite prognostic markers [25–30]. Such tools could help distinguish infants likely to benefit from KPE from those more appropriately managed with early transplant strategies. The key issue is therefore not PLT versus KPE in general, but which sequence and timing of surgery and transplantation best serve each individual child.

This study has several limitations. Its retrospective registry-based design resulted in some missing data, and subgroup

13993046, 2026, 10, Downloaded from https://onlinelibrary.wiley.com/doi/10.1111/petr.70458 by Universita Di Brescia, Wiley Online Library on [06/11/2026]. See the Terms and Conditions (https://onlinelibrary.wiley.com/terms-and-conditions) on Wiley Online Library for rules of use; OA articles are governed by the applicable Creative Commons License

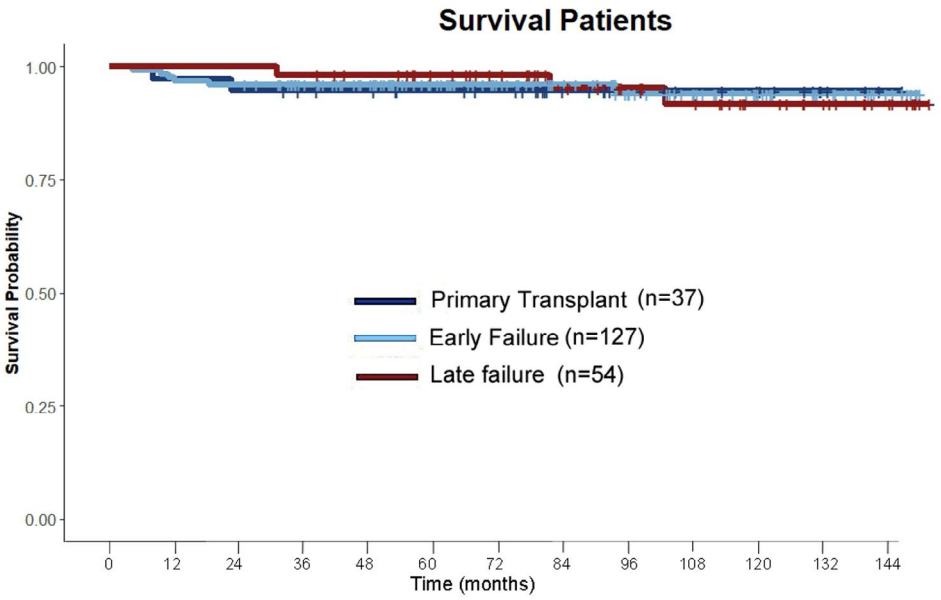


FIGURE 4 | Patient survival in a cohort of 218 Biliary Atresia children who underwent liver transplantation (Italy 2012–2022), according clinical indication: Primary liver transplantation (no Kasai portoenterostomy), early failure of Kasai or late failure after Kasai.

analyses were limited by sample size. In addition, the multicenter nature of the registry introduced heterogeneity in referral pathways, medical management, and transplant practice. However, this real-world heterogeneity also reflects everyday clinical practice and increases the general relevance of the findings.

5 | Conclusion

KPE should remain the recommended first-line treatment for most infants with BA, but its benefit is limited to only a proportion of patients. A sizeable subgroup proceeds rapidly to LT after early KPE failure or presents too late with advanced disease and is considered for PLT from the outset. Earlier identification of infants unlikely to benefit from KPE, prompt listing once a transplant-first strategy is chosen, and broader access to technical options for very small recipients (split, living-donor and ABO-incompatible LT) are the key elements to reduce pretransplant waiting and the associated clinical deterioration, and optimizing the BA management at a national level.

Author Contributions

Jean de Ville de Goyet: conceptualization. **Giovanni Boroni, Jean de Ville de Goyet, Nicolo Rizzitello:** methodology. **Giovanni Boroni, Jean de Ville de Goyet, Nicolo Rizzitello:** formal analysis. **Jean de Ville de Goyet, Giovanni Boroni, Giusy Ranucci:** writing – original draft. **Jean de Ville de Goyet, Giovanni Boroni, Giusy Ranucci, Daniela Liccardo, Daniele Alberti, Marco Spada, Valeria Casotti, Raffaele Iorio, Pier Luigi Calvo, Nicolo Rizzitello:** writing – review and editing. **Jean de Ville de Goyet, Giusy Ranucci:** supervision.

Acknowledgments

We thank the Italian Society of Pediatric Gastro-Enterology and Nutrition (SIGENP), l'Associazione Malattie Epatiche Infantili (AMEI) for their support and collaboration, BIOMEDIA, a national provider for supporting education and science, for their invaluable assistance in maintaining the Italian Biliary Atresia Registry and facilitating the research activities, and all the members of the working group "Network AVB" (https://www.avb-sigenp.org/site/home) (Table S1). Their contributions were essential to the successful completion of this work. The authors used Perplexity for 1-improving structure as per JPS guidelines, 2-language editing (to improve grammar, spelling, punctuation, and clarity), 3-shorten where appropriate and eliminating repetitions. AI was used exclusively at the final step of the manuscript preparation, when the data analysis had been finalized and when the discussion had been already drafted. The AI tool was demanded to not generate, add, or modify in any other manner the content or meaning of the text. No AI tools were used in the analysis of the data or writing of the initial manuscript. The authors reviewed the final text and any revisions made by AI software, and accept full responsibility for the manuscript's content.

Funding

The authors have nothing to report.

Conflicts of Interest

Any financial or personal relationships that could be perceived to influence the results are disclosed by each author separately, if any.

Data Availability Statement

The complete dataset underlying this article cannot be shared publicly because of privacy restrictions protecting participant confidentiality. Data may be shared on reasonable request to the corresponding author, subject to a formal data access agreement ensuring ethical compliance and anonymization; all associated costs will be borne by the requester.

References

1. C. Jimenez-Rivera, U. D. Ekong, and V. A. McLin, "International Incidence and Outcomes of Biliary Atresia: A Systematic Review of Population-Based Studies," *Journal of Pediatric Gastroenterology and Nutrition* 56 (2013): 676–682.

13993046, 2026, 10, Downloaded from https://onlinelibrary.wiley.com/doi/10.1111/petr.70458 by Universita Di Brescia, Wiley Online Library on [06/11/2026]. See the Terms and Conditions (https://onlinelibrary.wiley.com/terms-and-conditions) on Wiley Online Library for rules of use; OA articles are governed by the applicable Creative Commons License

2. M.-H. Chang, M.-W. Lai, C.-H. Hsiao, et al., "Reduction of the Ages at Diagnosis and Operation of Biliary Atresia in Taiwan: A 15-Year Population-Based Cohort Study," *World Journal of Gastroenterology* 21 (2015): 13080–13087.

3. P. J. McKiernan, A. J. Baker, C. Lloyd, G. Mieli-Vergani, and D. A. Kelly, "British Paediatric Surveillance Unit Study of Biliary Atresia: Outcome at 13 Years," *Journal of Pediatric Gastroenterology and Nutrition* 48 (2009): 78–81.

4. R. A. Schreiber, C. C. Barker, E. A. Roberts, et al., "Biliary Atresia: The Canadian Experience," *Journal of Pediatrics* 151 (2007): 659–665.e1.

5. G. Ranucci and D. Alberti, Establishing a SIGENP Italian Network for the Study of Biliary Atresia (2018).

6. F. Di Dato, G. Ranucci, J. de Ville de Goyet, D. Alberti, and R. Iorio, "Unusual Clinical Course for Untreated Malformative Biliary Atresia Infant: Is Portal Hypertension an Important Driver of Liver Fibrosis?," *Journal of Pediatric Gastroenterology and Nutrition* 72 (2021): 216–219.

7. M. Davenport, E. Ong, K. Sharif, et al., "Biliary Atresia in England and Wales: Results of Centralization and New Benchmark," *Journal of Pediatric Surgery* 46 (2011): 1689–1694.

8. M. Fanna, G. Masson, C. Capito, et al., "Management of Biliary Atresia in France 1986 to 2015: Long-Term Results," *Journal of Pediatric Gastroenterology and Nutrition* 69, no. 4 (2019): 416–424.

9. W. de Vries, Z. J. de Langen, H. Groen, et al., "Biliary Atresia in The Netherlands: Outcome of Patients Diagnosed Between 1987 and 2008," *Journal of Pediatrics* 160 (2012): 638–644.e2.

10. R. A. Schreiber, C. C. Barker, E. A. Roberts, S. R. Martin, and Canadian Pediatric Hepatology Research Group, "Biliary Atresia in Canada: The Effect of Centre Caseload Experience on Outcome," *Journal of Pediatric Gastroenterology and Nutrition* 51 (2010): 61–65.

11. B. E. Wildhaber, P. Majno, J. Mayr, et al., "Biliary Atresia: Swiss National Study, 1994-2004," *Journal of Pediatric Gastroenterology and Nutrition* 46 (2008): 299–307.

12. J. Leonhardt, J. F. Kuebler, P. J. Leute, et al., "Biliary Atresia: Lessons Learned From the Voluntary German Registry," *European Journal of Pediatric Surgery* 21 (2011): 82–87.

13. S. P. Alexopoulos, M. Merrill, C. Kin, et al., "The Impact of Hepatic Portoenterostomy on Liver Transplantation for the Treatment of Biliary Atresia: Early Failure Adversely Affects Outcome," *Pediatric Transplantation* 16 (2012): 373–378.

14. M. Davenport and R. Superina, "Primary Liver Transplant in Biliary Atresia: The Case for and Against," *Journal of Pediatric Surgery* 59 (2024): 1418–1426.

15. J. Zhan, S. Liu, T. Li, and X. Li, "Kasai Procedure or Liver Transplantation: How Should We Choose in Biliary Atresia?," *Hepatobiliary Surg Nutrition* 13 (2024): 1019–1021.

16. J. Seda Neto, J. S. Neto, F. H. Feier, et al., "Impact of Kasai Portoenterostomy on Liver Transplantation Outcomes: A Retrospective Cohort Study of 347 Children With Biliary Atresia," *Liver Transplantation* 21 (2015): 922–927.

17. R. Okubo, M. Nio, H. Sasaki, and M. Wada, "Japanese Biliary Atresia Registry," *World Journal of Pediatric Surgery* 8 (2025): e001024.

18. K. Uto, Y. Inomata, S. Sakamoto, T. Hibi, H. Sasaki, and M. Nio, "A Multicenter Study of Primary Liver Transplantation for Biliary Atresia in Japan," *Pediatric Surgery International* 35 (2019): 1223–1229.

19. E. LeeVan, L. Matsuoka, S. Cao, S. Groshen, and S. Alexopoulos, "Biliary-Enteric Drainage vs Primary Liver Transplant as Initial Treatment for Children With Biliary Atresia," *JAMA Surgery* 154 (2019): 26–32.

20. D. Yoeli, R. A. Choudhury, S. S. Sundaram, et al., "Primary vs. Salvage Liver Transplantation for Biliary Atresia: A Retrospective Cohort Study," *Journal of Pediatric Surgery* 57, no. 10 (2022): 407–413.

21. C. Chardot, M. Carton, N. Spire-Bendelac, et al., "Is the Kasai Operation Still Indicated in Children Older Than 3 Months Diagnosed With Biliary Atresia?," *Journal of Pediatrics* 138, no. 2 (2001): 224–228.

22. C. P. Lemoine, J. P. LeShock, K. A. Brandt, and R. Superina, "Primary Liver Transplantation vs. Transplant After Kasai Portoenterostomy for Infants With Biliary Atresia," *Journal of Clinical Medicine* 11 (2022): 3012.

23. M. Utz Melere, V. Sanha, M. Farina, et al., "Primary Liver Transplantation *vs* Transplant After Kasai Portoenterostomy in Children With Biliary Atresia: A Retrospective Brazilian Single-Center Cohort," *World Journal of Transplantation* 14, no. 1 (2024): 88734.

24. R. Tambucci, C. de Magnée, M. Szabo, et al., "Sequential Treatment of Biliary Atresia With Kasai Hepatoportoenterostomy and Liver Transplantation: Benefits, Risks, and Outcome in 393 Children," *Frontiers in Pediatrics* 9 (2021): 697581.

25. H. Tomita, N. Shimojima, H. Sasaki, et al., "Predicting Cirrhosis and Poor Outcomes of Bile Drainage Surgery for Biliary Atresia: A Multicentric Observational Study in Japan," *Annals of Surgery* 279 (2024): 692–698.

26. C. Ye and W. Gao, "Predictors of Outcome Among Children With Biliary Atresia: A Single Centre Trial," *PeerJ* 13 (2025): e19001.

27. L. Cui, Y. Xu, Y. Liu, et al., "Value of Preoperative APRI and FIB-4 in Assessing Short-Term Prognosis After Kasai Portoenterostomy," *World Journal of Pediatric Surgery* 8 (2025): e001002.

28. Q. Hua, Z. Ding, H. Xie, and W. Tang, "Correlation Analysis Between Preoperative Liver Ultrasound Elastography and Postoperative Jaundice Clearance After Kasai Surgery for Biliary Atresia," *Journal of Pediatric Surgery* 60 (2025): 162454.

29. R. M. Jeropoulos, J. Arroyo, and M. Davenport, "Predicting and Optimising Outcome for Biliary Atresia," *Seminars in Pediatric Surgery* 33 (2024): 151479.

30. L. Y. Sun, "Enhancing Prognostic Insights in Biliary Atresia: Implications of the iBALF Score," *HepatoBiliary Surgery and Nutrition* 14 (2025): 121–124.

Supporting Information

Additional supporting information can be found online in the Supporting Information section. **Table S1:** AVB network working group.

13993046, 2026, 10, Downloaded from https://onlinelibrary.wiley.com/doi/10.1111/petr.70458 by Universita Di Brescia, Wiley Online Library on [06/11/2026]. See the Terms and Conditions (https://onlinelibrary.wiley.com/terms-and-conditions) on Wiley Online Library for rules of use; OA articles are governed by the applicable Creative Commons License