Received: 25 March 2026 | Accepted: 4 June 2026

DOI: 10.1002/jpn3.70497

JPGN

ORIGINAL ARTICLE

The Italian biliary atresia registry: Insights and lessons from the retrospective analysis of a 10-year period cohort

Giovanni Boroni[1] | Daniela Liccardo[2] | Daniele Alberti[1] | Jean de Ville de Goyet[3,4] | Fabio Fusaro[5] | Marco Spada[6] | Valeria Casotti[7] | Mara Cananzi[8] | Giuseppe Indolfi[9] | Raffaele Iorio[10] | Pier Luigi Calvo[11] | Annalisa Madeo[12] | Francesco Cirillo[13] | Angelo Colucci[14] | Mara Colusso[15] | Francesco Fascetti Leon[8] | Elisa Bartolini[9] | Fabiola Di Dato[10] | Maurizio Cheli[15] | Claudia Mandato[16,17] | Angelo Di Giorgio[18] | Fabio Tuzzolino[19] | Giusy Ranucci[3,4] | for the Italian Society of Paediatric Gastroenterology, Hepatology, and Nutrition (SIGENP) AVB Network Working Group

Abstract

Objectives: To describe national patterns of management, outcomes, and prognostic factors in infants with biliary atresia (BA) in Italy.

Methods: This retrospective study used data from the Italian BA Registry from January 2012 to December 2021. Overall, 309 infants with BA were identified. Clearance of jaundice (CoJ) was defined as total bilirubin <1.2 mg/dL within 6 months. Univariate and multivariate analyses were used to identify prognostic factors for native liver and patient survival.

Results: Kasai portoenterostomy (KPE) was performed in 272 infants (88%) at a median age of 68.5 days (interquartile range 52–83). The CoJ rate after KPE was 39%. Multivariate analysis confirmed that CoJ was significantly associated with younger age and lower total bilirubin at referral ($p = 0.0446$ and $p = 0.0144$, respectively), younger age at KPE ($p = 0.0345$), and postoperative corticosteroid use ($p = 0.0036$). Multiple logistic regression confirmed the major effect of steroid use: the odds ratio for steroid use was 2.51 (95% confidence interval 1.38–4.58; $p = 0.0027$), whereas the odds ratio for age at KPE was 0.987 (95% CI: 0.975–0.999; $p = 0.0371$). Liver transplantation was performed in 212/309 patients, and overall 5-year actuarial patient survival was 97.4%.

Conclusions: This retrospective analysis confirms that age at KPE and postoperative corticosteroid use are key favorable factors for successful CoJ, with steroid use showing the strongest association. The excellent overall patient survival highlights the critical role of management in expert centers, even within a decentralized model.

KEYWORDS

Kasai portoenterostomy, native liver survival, neonatal diagnosis, pediatric liver transplantation, steroids

For affiliations refer to page 7.

Correspondence: Jean de Ville de Goyet, IRCCS ISMETT, ViaTricomi, Palermo, Italy. Email: Jdeville@Ismett.edu

Funding information: None

Giovanni Boroni and Daniela Liccardo equal first authorship.

Fabio Fusaro: Scientific Committee of SIGENP AVB Network Working Group.

Claudia Mandato and Angelo Di Giorgio: SIGENP representative.

Disclaimer: Although this paper is produced by the SIGENP - Biliary Atresia Network Working Group, it does not necessarily represent SIGENP policy and is not endorsed by SIGENP.

1 | INTRODUCTION

Kasai portoenterostomy (KPE) is the only currently available conservative treatment for patients with biliary atresia (BA). Its aim is to restore bile flow and prevent the otherwise inexorable progression to biliary cirrhosis and end-stage liver failure.[1–4] Large series and national registries have highlighted several prognostic factors that influence KPE success and long-term native liver survival (NLS).[5–16]

The Italian Biliary Atresia Network (IBAN) was established in March 2018 under the auspices of the Italian Society of Pediatric Gastroenterology, Hepatology, and Nutrition (SIGENP) (Table S1). This report presents the retrospective analysis of the initial dataset, covering the 10-year period from 2012 to 2021 in a decentralized care model. It provides insight into current BA management in Italy and identifies prognostic factors relevant to the national context.[17,18] Particular attention is given to the role of steroid therapy at KPE.

What is Known

- Successful Kasai operation in infants born with biliary atresia depends mostly on early diagnosis.
- The role of steroid therapy remains controversial.

What is New

- This analysis emphasizes the significant role of steroids on outcome—more impactful than age at surgery.
- Excellent patient outcome (>95% at 5 years) can be achieved.
- A decentralized care system can meet the challenge of offering adequate outcomes.

2 | METHODS

2.1 | Ethics statement

The study was approved by the SIGENP scientific board (NP3854), the National Italian Ethics Committee, and the ethics committee of each participating center. The study protocol conformed to the ethical guidelines of the 1975 Declaration of Helsinki, as reflected by approval from the research and ethics committees of all participating institutions.

Informed consent was obtained prospectively from the parents or legal guardians of all patients included in this study. In accordance with the protocol governing the national registry, written informed consent was specifically required from each family prior to the inclusion of their child's data in the registry. This consent explicitly covered the retrospective use of registry data for clinical research purposes and scientific publication.

2.2 | Study design

This retrospective study was based on the BA Registry and covered the period from January 2012 to December 2021. All measures were taken to ensure capture of all BA patients diagnosed in Italy during the study period. Details of participating centers are reported in Table S1. Enrollment was based on diagnosis, and many patients enrolled by one center were operated on in another. Seven of 13 centers performed KPE, including five that also performed transplantation. Therefore, the concept of "center" in this study was not based on KPE performance, and no analysis related to KPE surgical expertise was performed.

2.3 | Patients selection

Patients registered in the Registry were enrolled in the study. Collected data included general information, preoperative and perioperative clinical details, anatomical and histological findings, and follow-up information.

2.4 | Endpoints

- The process of care after BA diagnosis, and the general outcome, were analyzed with endpoints being,
  1. achieving Kasai operation, or not
  2. the survival with native liver
  3. the need for liver transplantation (LT), or death before LT
  4. post-transplantation outcome (death or survival)
  5. the overall patient survival (regardless of the type of care).
- The KPE outcome and the effect of steroid therapy after KPE, were assessed in terms of:
  1. clearance of jaundice (CoJ) (defined as total bilirubin ≤1.2 mg/dL (≤20 μmol/L) within 6 months after KPE).[3,19,20]
  2. NLS, or death or the need for LT.
- The outcomes of patients who did not undergo KPE were assessed by:
  1. death before LT or achieving LT
  2. post-transplantation outcome (death or survival).

2.5 | Statistical analysis

For descriptive and exploratory analyses, categorical variables were summarized as absolute and relative frequencies (counts and percentages), whereas continuous variables were expressed as median and interquartile range (IQR). Group comparisons were performed using the *T*-test for independent samples or the Wilcoxon rank-sum test for continuous variables, as appropriate, and the χ^2 test or Fisher's exact test for categorical variables. Survival probabilities were estimated using the Kaplan–Meier method, and differences between strata were assessed by the log-rank test. Patients alive without liver transplantation at the time of data extraction were considered censored.

Univariate and multivariate analyses were conducted to identify significant prognostic factors. Both univariate and multivariate logistic regression analyses were performed for binary outcomes, whereas time-to-event outcomes were investigated using Cox proportional hazards regression models. In multivariate analyses, significant predictors were identified through a backward selection procedure.

The list of primary and secondary/associated outcome variables used for statistical analyses is reported in Table S2.

3 | RESULTS

3.1 | Patients characteristics

The Registry identified 309 infants. Based on live birth data from the Italian National Institute of Statistics (Istat; www.istat.it), the calculated incidence in Italy during the study period was 1/15,142 live births, which is higher than that reported in some other European countries (the United Kingdom, 1/17,000; France, 1/19,600; the Netherlands, 1/18,619; Switzerland, 1/17,800).

The median age at referral was 58 days (IQR 37–77), with 14 infants diagnosed after 120 days of life (Figure S1). Median birth weight was 3.15 kg (IQR 2.8–3.4 kg), and median gestational age was 39 weeks (IQR 38–40). Among the 309 cases, 34 infants (11%) presented with biliary atresia splenic malformation (BASM) syndrome.

KPE was performed in 272 infants (88%). In this group, the median age at referral was 55 days (IQR 35–72), and the median age at KPE was 68.5 days (IQR 52–83).

3.2 | CoJ after KPE

CoJ was achieved in 39% of cases. When CoJ was achieved, NLS at 1 and 5 years after KPE was 94% and 84%, respectively. When CoJ was not achieved, NLS at 1 and 5 years was 44% and 7%, respectively ($p < 0.0001$) (Figure 1, left).

Univariate and multivariate analyzed identified factors associated with a higher CoJ rate.

a. Lower total bilirubin at referral ($p = 0.0144$).
b. Younger age at KPE ($p = 0.0345$).
c. Postoperative corticosteroid use ($p = 0.0036$).
d. BASM condition, AST or ALT or GGT levels at referral did not reach significance as prognostic parameters for CoJ.

3.3 | NLS

After KPE, overall actuarial NLS rates at 1, 2, and 5 years were 57.9%, 40.8%, and 32.5%, respectively (Figure 1, right). Median follow-up was 7.2 years (Q1–Q3: 4.8–10.0).

Uni- and multivariate analysis showed that a higher NLS rate was associated with:

a. Younger age at referral ($p = 0.0115$) (odds ratio [OR]: 1.014, 95% confidence interval [CI]: 1.003–1.024).
b. Lower total bilirubin at referral ($p = 0.0254$) (OR: 1.107, 95% CI: 1.013–1.211).
c. Younger age at KPE ($p = 0.0165$) (OR: 1.015, 95% CI: 1.003–1.027).
d. Postoperative corticosteroid use ($p = 0.0212$) (OR: 0.492, 95% CI: 0.269–0.899).
e. CoJ at 6 months post KPE ($p < 0.001$) (OR: 0.017, 95% CI: 0.007–0.038).
f. Post-KPE cholangitis, or BASM condition did not reach significance as prognostic parameters for NLS.

Analyzing the effect of age at KPE by comparing subgroups of patient (defined by conventional cut-off age values[5,9,18,19]) confirmed that >90 days age at KPE was a significant risk factor compared with KPE before the age of 45 days of life ($p = 0.0062$) (Figure 2).

In the time-dependent Cox model, the effect of age at KPE on NLS varied over time. Older age at KPE had a stronger detrimental impact during the early post-KPE period, with progressive attenuation thereafter. In other words, older age at KPE significantly increased early native liver loss, whereas late losses were not significantly associated with age at KPE. Consistently, the proportional hazards assumption was violated ($p = 0.021$), supporting inclusion of a time-dependent effect (Figure S2).

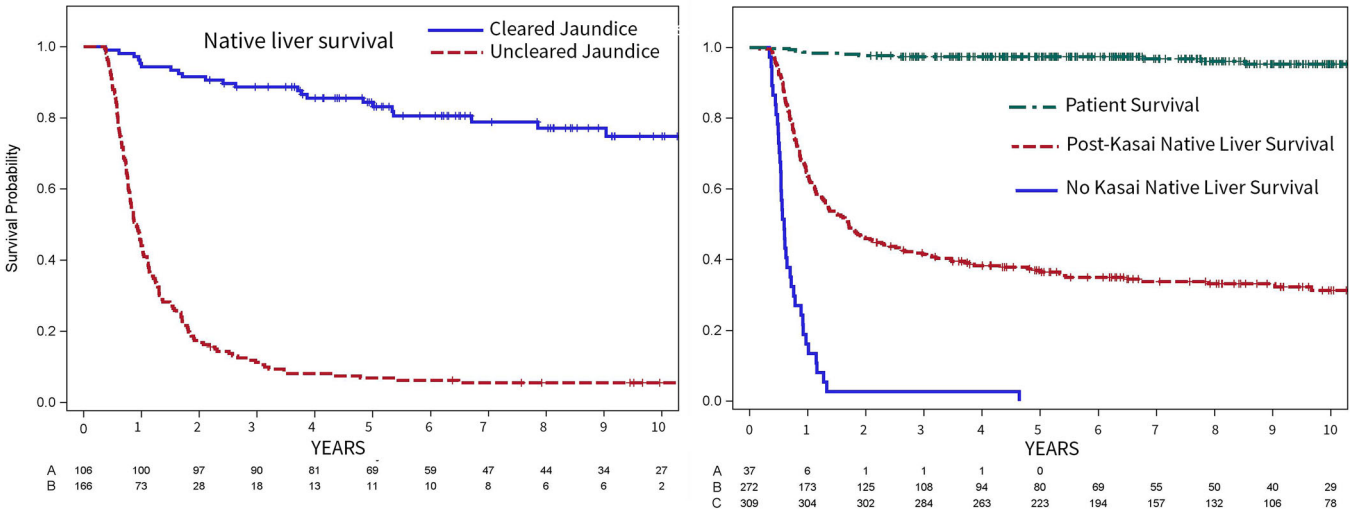

FIGURE 1 Overall patient and native liver survival. Left: Native liver survival after KPE according to whether CoJ was achieved. CoJ was defined as serum total/conjugated bilirubin <20 μmol/L (<1.2 mg/dL) within 6 months after KPE ($p < 0.0001$). Right: Overall Kaplan–Meier survival for patients (upper line) and for native liver after KPE (middle line) or primary transplantation (lower line) ($p < 0.0001$). CoJ, clearance of jaundice; KPE, Kasai portoenterostomy.

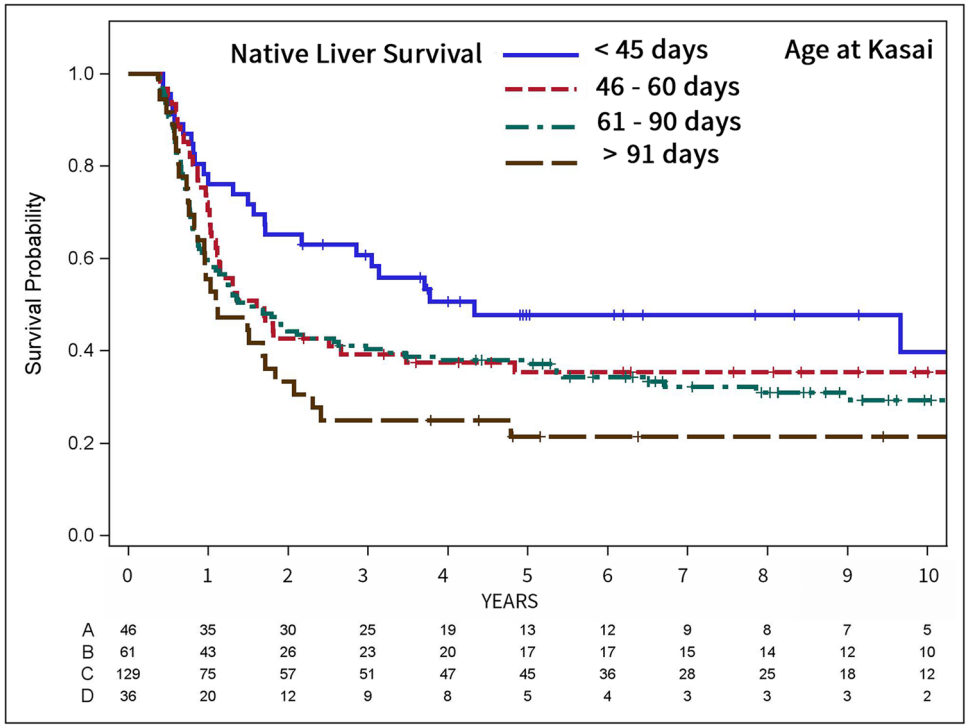

FIGURE 2 Native liver survival after KPE according to age at KPE. Age at surgery: A: <45 days; B: 46–60 days; C: 61–90 days; D: >91 days.[5,9,18,19] (Overall $p = 0.0765$; A vs. C, $p = 0.0062$). KPE, Kasai portoenterostomy.

3.4 | Effect of steroid therapy at/after KPE

Steroid use varied between and within enrolling centers (Figure S3), with the same variation observed between centers performing Kasai. It was scattered throughout the study period in a pattern close to, although not equivalent to, true randomization (Figure S4). Steroid protocols are described in Figure S5. Six of seven centers used protocol A, and one center used protocol B. Variations between centers mainly concerned the second and third months after KPE and were mostly related to early tapering in cases with obvious KPE failure.

Steroids were used in 71% of KPE cases. Overall, steroid use at KPE had a significant favorable effect on outcome, with increased NLS ($p = 0.0032$) (Figure 3, left). In univariate and multivariate analyses, no general patient parameter was significantly associated with steroid use; in particular, age at referral and age at KPE were homogeneously distributed. However, steroid use differed significantly between expert centers ($p < 0.0001$). Among children who eventually lost their liver (Figure 3, right), steroid therapy was not detrimental and was associated with a non-significant trend toward delayed need for LT ($p = 0.14$).

A more detailed "intended-CoJ" analysis showed that steroid use after KPE had a major effect, even greater than age at KPE itself. In multiple logistic regression, the odds ratio for steroid use was 2.51 (95% CI: 1.2–4.2; $p = 0.011$), whereas the odds ratio for age at KPE was 0.99 (95% CI: 0.97–1.0; $p = 0.0371$). In other words, KPE success decreased progressively with age at operation, with each additional day reducing the odds of CoJ by approximately 1.3%; this effect was independent of steroid use. Steroid therapy was associated with a significantly higher chance of CoJ at any given age at KPE and increased the chance of CoJ by approximately 2.5-fold.

3.5 | Death after KPE, or rescue by liver transplantation

Of the 272 patients who had KPE, 176 were listed later for LT and 175 (64.3%) have undergone LT (no patient was on waiting list at the time of writing) (Figure S1). Age at waiting list inscription and at LT were 8.6 (IQR 6.2–15.0) and 11.5 (IQR 7.8–20.5) months, respectively. Waiting time was 45 days (IQR 22–106) days, and pediatric end-stage liver disease (PELD) score at registration on waiting list was 18 (IQR 11–22). Thirty-seven other infants (12%) were managed by primary LT.

Five KPE children have died before LT. One child died at 18.5 months of age, who had been registered for LT at age 14 months: he died during an acute episode of hemorrhage before LT. Four other children died within their first year of life; deaths were related to prematurity with chromosomal abnormalities or multiple anomaly syndrome, acute hemorrhage, and RSV infection (1 case each).

3.6 | Overall patient survival

Among 309 infants, 92 survived with their native liver and 206 were successfully transplanted (Figures 1 and S5). No patient died before being proposed for either KPE or primary LT. Of the 175 children proposed for LT after KPE, 9 died: 5 before LT and 4 after LT (5%). In the whole cohort, 11 patients died: 5 after KPE and 6 after LT (2 primary LT, 4 post-KPE LT) (Figure 1).

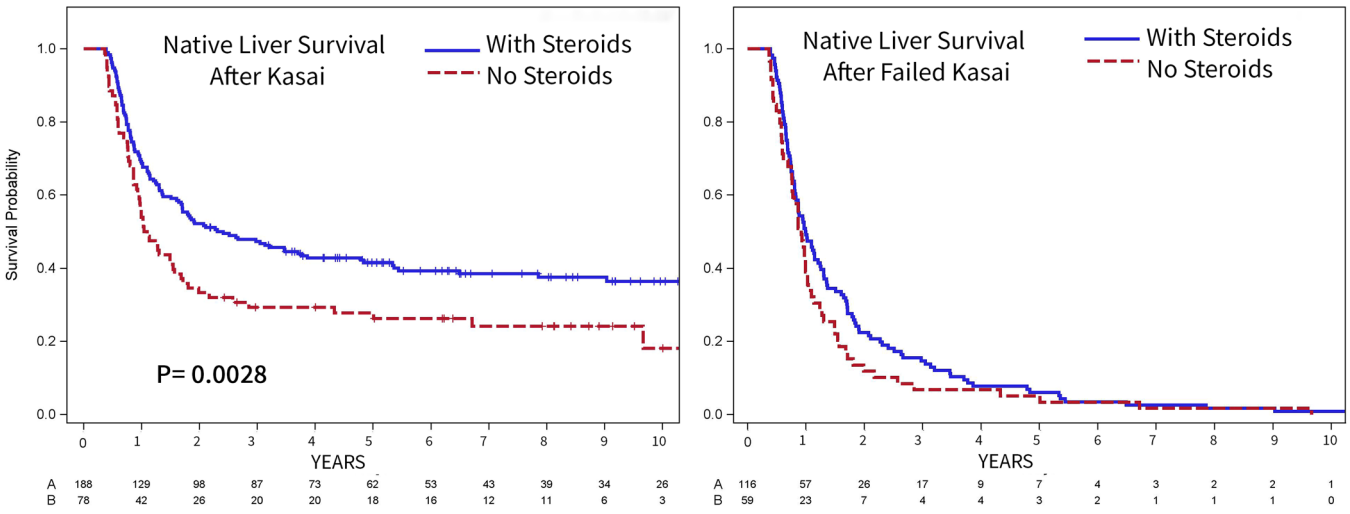

FIGURE 3 Native liver survival according to steroid use after KPE in infants with BA. Left: Overall native liver survival after KPE with or without steroids, showing a significantly favorable effect when steroids were used at KPE ($p = 0.0028$). Right: Dynamics of native liver loss among patients who lost the liver, with or without steroid use after KPE. Steroid use in the absence of bile-flow restoration was not detrimental. Although steroid use delayed the need for LT, this effect was not significant ($p = 0.14$). BA, biliary atresia; KPE, Kasai portoenterostomy; LT, liver transplantation.

No care pathway—successful KPE, LT after KPE, or primary LT—was significantly better or worse than another in terms of protecting the life of a child with BA ($p = 0.23$) (Figure S6).

3.7 | Center effect

Thirteen centers entered data into the Registry, with caseloads ranging from 1 to 98 patients.

There were no significant differences between centers in patient characteristics; in particular, age at referral and age at KPE were similar (Figures S7 and S8). There were significant differences in CoJ rate and NLS between centers ($p < 0.001$ and $p < 0.0001$, respectively). These differences were secondary to highly significant differences in steroid use at KPE ($p < 0.0001$). In other words, centers that did not use steroids had worse outcomes.

4 | DISCUSSION

In modern management of BA, the key prognostic factors for a good long-term outcome after KPE alone, defined as NLS, are early diagnosis of BA[2,10] and KPE performed at a young age.[9,19] Referral for LT is a third essential element for patient survival, but it also marks the end of NLS.[2,19,20] From this perspective, LT may be viewed as a failure of primary management, although it remains life-saving for many patients with BA. Against this background, several important messages emerge from this study.

First, the age at referral for suspected BA was high in this cohort (58 days, IQR 37–77 days), although it remained consistent with that reported in many other registries[20–24] (Table 1). Late referral also aligns with the unusually high proportion of patients proposed for primary LT in this cohort. These findings confirm that substantial work remains to be done in Italy to improve early suspicion, referral, and diagnosis. They may also explain the slightly lower CoJ rates observed after KPE compared with other registries (Table 1). Interestingly, the NLS in this cohort was broadly comparable with the mean NLS reported elsewhere[19,21,22] (Table 1). This observation may suggest that, within the age range at KPE commonly reported in most registries (50–70 days), moderate differences in age at KPE may not substantially affect overall outcomes. One possible explanation is that once cirrhosis is established, as occurs with late diagnosis and late KPE, CoJ may no longer be a sufficiently reliable prognostic marker for very long-term outcome.[25,26]

Importantly, the 5-year patient survival of 96.4% in this series compares favorably with published data[1] (Table 1). LT was performed in a large proportion of children in this series, approximately two-thirds of patients, and with a very high success rate (95% survival after primary LT). This may suggest that the timing of LT was appropriate, allowing elective transplantation in patients who were still in good clinical condition. This aspect warrants further study, although the registry cannot currently explore these issues in detail. The excellent overall patient survival in this cohort confirms the important role of LT in BA management. At some point in the clinical course of a patient with BA, it may be more important to focus on survival and quality of life than to avoid or delay LT at all costs.[19,21,22] This concept is reinforced by the similar mortality rates observed before and after LT in this series (2.2% and 1.8%, respectively).

Second, the analysis supports previous findings showing that earlier diagnosis and very early KPE significantly increase the likelihood of successful KPE and achievement of CoJ, the best indicator of effective bile-flow restoration. Among all variables studied, steroid use after KPE was an important and statistically significant favorable factor for CoJ. Evidence supporting the benefit of steroids after KPE has accumulated over the past two decades.[6,13,14] Its significance in the present study is notable because steroids were used non-randomly and without a standardized protocol, a setting that would usually be expected to produce more contradictory

TABLE 1 Outcomes from published biliary atresia multicentric cohorts and registries.

Country	First author (Year)	*N* Cases	Study period	Age at Kasai (days)	BASM (%)	Jaundice clearance (%)	Native liver survival	Primary liver transplant (%)	Overall patient long-term survival
Japan	Nio M (2003)	1381	1989–1999	-	2	62	10-year (1989 series) 53%	0.15	1989 series 5-year 75%. 10 year 69.4%
Switzerland	Wildhaber BE (2008)	48	1994–2004	68	8	39	2-year 43%	11.6	2-year 91%
England	Davenport M (2011)	439	1999–2009	54	12	55	5-year 46%	3.4	5-year 90%
United States	Shneider BL (2006)	104	1997–2000	61	14	40	2 year 56%	-	2 year 91%
Canada	Schreiber RA (2007)	349	1985–2002	65	-	-	4-year 33% 10-year 24%	11	4-year 79% 10-year 77%
The Netherlands	De Vries W (2012)	210	1998–2008	59	-	38	5-year 42% 10-year 27%	2.8	5-year 76%
France	Chardot C (2013)	1107	1986–2009	59	8	38	5-year 40% 10-year 36%	4.2	5-year 89% 20-year 77%
Nordic countries	Lampela H (2018)	154	2005–2016	64	12	64	5-year 53% 10-year 45%	3.9	5-year 88% 10year 87%
France	Fanna M (2019)	1428	1986–2015	59	11	39	5-year 41% 10-year 35%	4.8	5-year 87% (2010-2015)
Saudi Arabia	Abanemai, M (2022)	204	2000–2018	70	11	45	5-year 27%	25	10-year 72%
Germany	Madadi-Sanjani O (2022)	173	2010–2014	-	-	-	2-year 29%	7.5	2-year 88%
Japan	Okubo R (2025)	3951	1989–2023	65	-	63	10-year 50%	-	10-year 89%
England	Davenport M (2025)	867	1999–2019	51	13	61	5-year 51% 10-year 46%	21.8	5-year 91% 10-year 90%
THIS SERIES	Boroni G (2025)	309	2012– 2021	69	12	38	2-year 45% 5-year 33%	12.0	5-year 97% 10-year 95%

Abbreviation: BASM, biliary atresia splenic malformation.

results. Thus, this finding may be statistically meaningful despite the non-protocolized use of steroids.

Third, overall patient survival was excellent: 97% at 5 years and 95% at 10 years, comparing favorably with existing registries and large series[1] (Table 1). Survival among children diagnosed with BA during the study period was independent of the clinical strategy or care pathway. Patients proposed for primary LT, those with successful KPE, and those who underwent sequential KPE followed by LT had similar chances of surviving an otherwise lethal condition. This highlights the life-saving impact of LT within an expert network, while also emphasizing that native liver outcomes remain strongly dependent on early diagnosis and successful KPE. These findings confirm the known benefit for children with BA of being managed early within a BA-expert center, whether in a centralized or decentralized model.[21,27–31] As previously noted by Davenport et al., the benefit may simply be that close multidisciplinary specialist care (rather than only surgical expertise) allows for optimization of care, and efficient preparation/optimal timing of transplantation - thereby achieving no mortality before LT and low mortality after LT.[7,21,27,28] These observations emphasize the need for early referral to an expert multidisciplinary center for infants with early KPE failure, and those who are candidates for primary LT.

Fourth, this analysis contributes further to the controversy surrounding the effect of steroids. It confirms that steroid use after KPE is associated with an increased CoJ rate (Figure S2). The effect of steroid therapy was strongly associated with CoJ, suggesting that steroids may act synergistically with KPE by amplifying bile-flow restoration. No steroid effect was observed in terms of liver protection when CoJ was not restored, and the native liver was eventually lost (Figure S2). When the objective was to achieve CoJ, the impact of steroid use after KPE was substantial and appeared even greater than the effect of age at KPE itself. In multiple logistic regression, the odds ratio for steroid use was 2.51 (95% CI: 1.2–4.2; $p = 0.011$), whereas the odds ratio for age at KPE was 0.99 (95% CI: 0.97–1.0; $p = 0.05$). In other words, at any given age at KPE, steroid therapy was associated with a significantly higher chance of achieving CoJ.

Finally, the aim of KPE is to restore bile flow, clear jaundice, slow liver damage, and prolong NLS. International data consistently highlight age at KPE as a critical determinant of success and outcome. Despite decades of calls for earlier diagnosis and earlier KPE, little progress has been made in achieving earlier access to surgery[31] and calls to improve this situation are now emerging worldwide. The evidence suggests that fibrosis is already advanced when KPE is performed at around 60 days of life and is likely to persist for life even when CoJ is achieved. Cirrhosis and portal hypertension remain common in long-term survivors, who may face severe comorbidities. Future standards should therefore aim for KPE closer to, or before, 30 days of age, rather than accepting the current usual timing of 50–70 days achieved worldwide (Table 1). Recent literature has shown superior outcomes with neonatal KPE, defined as KPE performed before 30 days of age.[7,8]

This study has several limitations. The first is its retrospective design, with incomplete data collection that led to the exclusion of several variables for which information was missing in more than 10% of cases, including BA type, albumin level, and platelet count after KPE. However, the most important parameters were analyzed. Second, although the series is not small, the cohort size limited the statistical analyses mainly to the overall group, and subgroup analyses were restricted to preserve statistical power. Third, as this was a registry study, data were contributed by many centers, creating heterogeneity in clinical care. However, this heterogeneity may also be considered a strength, as it introduced variation that may have mimicked a form of chance balance - thereby reinforced the statistical power of an otherwise not-randomized study (Figure S4).

5 | CONCLUSION

In summary:

- The success of KPE decreases progressively with age at operation, with each additional day reducing the odds of CoJ by approximately 1.3%.
- Steroid therapy was associated with a significantly higher chance of achieving CoJ at any given age at KPE, with an approximately 2.5-fold effect.

This inaugural retrospective analysis from the IBAN, including 309 patients diagnosed over a decade, provides important foundational data on the national management of BA. Although it showed that diagnosis was relatively late, that 12% of infants were not proposed for KPE, and that long-term NLS was relatively low, it also demonstrated that a decentralized model involving multiple expert centers achieved excellent overall patient survival. The study confirms that early diagnosis and early KPE provide the best outcomes, while also showing very satisfactory results in patients with late referral and those undergoing primary LT.

The next phase for IBAN will be the implementation of prospective national data collection. This process should facilitate multicenter research in Italy and represents a necessary step forward in the study of this very rare disease.

AFFILIATIONS

[1]UOC Chirurgia Pediatrica, Spedali Civili di Brescia, Brescia, Italy

[2]Division of Hepatology and Metabolic Diseases, Liver Transplant Unit, Bambino Gesù Children's Hospital, IRCCS, Rome, Italy

[3]Paediatric Liver Unit, ISMETT, Palermo, Italy

[4]UPMC Italy, Palermo, Italy

[5]Fetal and Neonatal Surgery Unit, Bambino Gesù Children's Hospital, IRCCS, Rome, Italy

[6]Division of Hepatobiliopancreatic Surgery, Liver and Kidney Transplantation, Bambino Gesù Children's Hospital, IRCCS, Rome, Italy

[7]Hepatology, Gastroenterology and Transplant Unit, ASST Papa Giovanni XXIII, Bergamo, Italy

[8]Gastroenterologia, Endoscopia Digestiva, Epatologia e Cura del Bambino con Trapianto di Fegato, Dipartimento di Salute della Donna e del Bambino, A.O.U. di Padova, Padova, Italy

[9]Epatologia Pediatrica Azienda Ospedaliero-Universitaria Meyer, Firenze, Italy

[10]UOS Epatologia Pediatrica, Università di Napoli Federico II, Napoli, Italy

[11]Gastroenterologia Pediatrica. Ospedale Infantile Regina Margherita, Azienda Ospedaliero-Universitaria Città della Salute e della Scienza di Torino, Torino, Italy

[12]UOC. di Gastroenterlogia ed Endoscopia digestiva, IRCCS Istituto Giannina Gaslini, Genova, Italy

[13]Pediatrics of Chronic Diseases, Hepatology and Nutrition - Santobono-Pausilipon Children's Hospital, Naples, Italy

[14]UOSD Pediatria e Assistenza Neonatale - Azienda Ospedaliera Universitaria "San Giovanni di Dio e Ruggi D'Aragona" - P.O. di Cava de' Tirreni, Salerno, Italy

[15]Chirurgia Pediatrica, Ospedale Papa Giovanni XIII, Bergamo, Italy

[16]Department of Medicine, Surgery and Dentistry University of Salerno, Italy

[17]Santobono-Pausilipon Children Hospital, Naples, Italy

[18]Pediatric Liver Service, Hospital Santa Maria Misericordia, University of Udine, Italy

[19]Statistics and Data Management Services, ISMETT, Palermo, Italy

ACKNOWLEDGMENTS

We thank the Italian Society of Pediatric Gastro-Enterology and Nutrition (SIGENP, https://www.avb-sigenp.org/site/home), l'Associazione Malattie Epatiche Infantili (AMEI) for their support and collaboration, and 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. Their contributions were essential to the successful completion of this work. We thank all the members of the Italian Biliary Atresia Network created in 2018, who supported this project and directly contributed to the creation of the Registry: Daniele Alberti, Roberto Antonucci, Elisa Bartolini, Pietro Betalli, Giovanni Boroni, Pierluigi Calvo, Mara Cananzi, Valeria Casotti, Francesco Cirillo, Maria Grazia Clemente, Angelo Colucci, Lorenzo D'Antiga, Jean de Ville de Goyet, Fabiola Di Dato, Angelo di Giorgio, Diletta Dona', Francesco Fascetti Leon, Fabio Fusaro, Giovanni Gaglione, Paola Gaio, Fabrizio Gennari, Giuseppe Indolfi, Raffaele Iorio, Alessandro La Rosa, Daniela Liccardo, Annalisa Madeo, Giuseppe Maggiore, Claudia Mandato, Caterina Marcato, Alessandra Nardi, Filippo Parolini, Michele Pinon, Giusy Ranucci, Francesca Sbravati, Marco Sciveres, Marco Spada, Maurizio Cheli, Mara Colusso, Davide Cintorino (Table S1). The authors used Perplexity exclusively for language editing to improve grammar, spelling, punctuation, and clarity, only at the final step of manuscript preparation. The 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 analysis of the data, writing of the initial manuscript or drafting of tables and figures. The authors reviewed the final text and any revisions made by AI software, and accept full responsibility for the manuscript's content. The Italian Biliary Atresia Network Working Group is an independent, self-supporting association of physicians with a special interest in the management of Biliary Atresia, covering a range of medical specialties. This working group has not received any specific funding for its general activities, including the preparation of this manuscript. The Italian Biliary Atresia Registry is owned by the Italian Society of Paediatric Gastroenterology, Hepatology, and Nutrition (SIGENP), while it is managed by BioMedia, a national provider of services for physicians and medical societies (including CME, administrative, and organizational support), with Teras MM Srl responsible for IT and technical aspects. BioMedia operates independently for the management and operation of the Registry. SIGENP independently manages any donations made in support of the Registry or the Network and allocates them to specific projects necessary for their functioning (e.g., biannual meetings, data management activities).

CONFLICT OF INTEREST STATEMENT

Although the Italian Registry for Biliary Atresia is supported financially by the Italian Society of Pediatric Gastro-Enterology and Nutrition (SIGENP), this research project was not specifically funded. The authors declare no conflicts of interest.

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

ORCID

Jean de Ville de Goyet https://orcid.org/0000-0001-7681-6178

REFERENCES

1. Jimenez-Rivera C, Ekong UD, McLin VA. International incidence and outcomes of biliary atresia: a systematic review of population-based studies. *J Pediatr Gastroenterol Nutr*. 2013;56:676-682.
2. Lin JS. Reduction of the ages at diagnosis and operation of biliary atresia in Taiwan: a 15-year population-based cohort study. *World J Gastroenterol*. 2015;21:13080-13087.
3. McKiernan P, Baker AJ, Lloyd C, Mieli-Vergani G, Kelly DA. British paediatric surveillance unit study of biliary atresia: outcome at 13 years. *J Pediatr Gastroenterol Nutr*. 2009;48:78-81.
4. Schreiber RA, Barker CC, Roberts EA, Martin SR, Dunn SP, Ng VL. Biliary atresia: the Canadian experience. *J Pediatr*. 2007;151:659-665.
5. Okubo R, Nio M, Sasaki H, Wada M. Japanese biliary atresia registry. *World J Pediatr Surg*. 2025;8:e001024.
6. Tabak F, Kurtaran B, Zerdali E, et al. Direct acting antivirals are effective and safe in intravenous drug users with chronic HCV preview of your abstract file infection: first report from Turkey. *Turkish J Gastroenterol*. 2019;30:61-62.
7. Davenport M, Parsons C, Tizzard S, Hadzic N. Steroids in biliary atresia: single surgeon, single centre, prospective study. *J Hepatol*. 2013;59:1054-1058.
8. Davenport M, Makin E, Ong EG, Sharif K, Dawrant M, Alizai N. The outcome of a centralization program in biliary atresia: twenty years and beyond. *Ann Surg*. 2025;281:608-614.
9. Yang C, Ke M, Zhou Y, Xu H, Diao M, Li L. Impact of early Kasai portoenterostomy on short-term outcomes of biliary atresia: a systematic review and meta-analysis. *Front Surg*. 2022;9:924506.
10. Goldstein SD, Lee LC, Kim Y, Hong JC, Perrone J. Guidance for the primary care provider in identifying infants with biliary atresia by 2-4 weeks of life: clinical report. *Pediatrics*. 2025;155:e2024070077.
11. Jain M, Sharma S, Singh A, et al. Incidence and prognostic factors associated with biliary atresia in western India. *Ann Hepatol*. 2019;18:573-579.
12. Sanghai SR, Shah I, Bhatnagar S, Murthy A. Incidence and prognostic factors associated with biliary atresia in western India. *Ann Hepatol*. 2009;8(2):120-122.
13. Tyraskis A, Parsons C, Davenport M. Glucocorticosteroids for infants with biliary atresia following Kasai portoenterostomy. *Cochrane Database Syst Rev*. 2018;5:CD008735.
14. Egozi EJ, Shaul DB, Dunn JC. Improved outcome of biliary atresia with postoperative high-dose steroid. *J Pediatr Surg*. 2006;41:855-858.
15. Shneider BL, Brown MB, Haber B, et al. A multicenter study of the outcome of biliary atresia in the United States, 1997 to 2000. *J Pediatr*. 2006;148:467-474.e1.
16. Chen Y, Li L, Chen J, Wen J, Wang S, Shen Z. Incidence and outcome of biliary atresia in Shanghai, China from 2015 to 2016: a cohort study. *BMJ Paediatr Open*. 2024;8(1):e002475.
17. Ranucci G, Alberti D, et al. *Establishing a SIGENP Italian Network for the Study of Biliary Atresia*. Research Gate; 2018. https://www.researchgate.

net/publication/328001787_P028_Establishing_a_SIGENP_Italian_network_for_the_study_of_biliary_atresia

18. Chardot C, Buet C, Serinet MO, et al. Improving outcomes of biliary atresia: French national series 1986–2009. *J Hepatol*. 2013;58:1209-1217.
19. de Vries W, de Langen ZJ, Groen H, et al. Biliary atresia in the Netherlands: outcome of patients diagnosed between 1987 and 2008. *J Pediatr*. 2012;160:638-644.e2.
20. AMEI. Il primo Network italiano per lo studio dell'Atresia delle Vie Biliari. https://www.ameiitalia.org/amei-progetti/avb-il-primo-network-italiano-per-lo-studio-dellatresia-delle-vie-biliari/
21. Davenport M, Superina R. Primary liver transplant in biliary atresia: the case for and against. *J Pediatr Surg*. 2024;59(8):1418-1426.
22. Al-Hussaini A, Abanemai M, Alhebbi H, et al. The epidemiology and outcome of biliary atresia: Saudi Arabian National Study (2000-2018). *Front Pediatr*. 2022;10:921948.
23. D'Antiga L, Vajro P, Cunsolo T, et al. Italian Cooperative Study Group on biliary atresia. Biliary atresia: 20-40-year follow-up with native liver in an Italian centre. *Dig Liver Dis*. 2019;51:575-579.
24. Nio M, Ohi R, Miyano T, Saeki M, Shiraki K, Tanaka K. Five- and 10-year survival rates after surgery for biliary atresia: a report from the Japanese biliary atresia registry. *J Pediatr Surg*. 2003;38:997-1000.
25. Zhan J, Liu S, Li T, Li X. Kasai procedure or liver transplantation: how should we choose in biliary atresia? *Hepatobiliary Surg Nutr*. 2024;13:1019-1021.
26. Shen WJ, Chen G, Wang M, Zheng S. Liver fibrosis in biliary atresia. *World J Pediatr*. 2019;15(2):117-123.
27. Davenport M, Ong E, Sharif K, et al. Biliary atresia in England and Wales: results of centralization and new benchmark. *J Pediatr Surg*. 2011;46:1689-1694.
28. Davenport M, De Ville de Goyet J, Stringer M, et al. Seamless management of biliary atresia in England and Wales (1999-2002). *Lancet*. 2004;363:1354-1357.
29. Lampela H, Ritvanen A, Kosola S, et al. National centralization of biliary atresia care to an assigned multidisciplinary team provides high-quality outcomes. *Scand J Gastroenterol*. 2012;47:99-107.
30. Serinet MO, Broué P, Jacquemin E, et al. Management of patients with biliary atresia in France: results of a decentralized policy 1986-2002. *Hepatology*. 2006;44:75-84.
31. Madadi-Sanjani O, Fortmann D, Rolle U, et al. Centralization of biliary atresia: has germany learned its lessons? *Eur J Pediatr Surg*. 2022;32:233-239.

SUPPORTING INFORMATION

Additional supporting information can be found online in the Supporting Information section at the end of this article.

How to cite this article: Boroni G, Liccardo D, Alberti D, et al. The Italian biliary atresia registry: Insights and lessons from the retrospective analysis of a 10-year period cohort. *J Pediatr Gastroenterol Nutr*. 2026;1-9. https://doi.org/10.1002/rhc3.70057.