CASE REPORT

Biliary atresia: variability in evolution after Kasai portoenterostomy. Report of two cases

Kerly Fiestas [1], Wilma Geraige [1], Mariana Torres [1], Paula Roitman [1], Karina Arco [1], Giuliana Vaquer [1]

[1] Pediatric Gastroenterology Service, Dr. Humberto Notti Hospital, Mendoza M5500, Argentina.

ABSTRACT

Background: Biliary atresia (BA) is a leading cause of neonatal cholestasis and the primary indication for liver transplantation in children. Its etiology is unknown and is characterized by the obliteration of the bile ducts, resulting in impaired bile flow, which manifests as jaundice, dark urine, pale stools, hepatomegaly, and/or splenomegaly. Kasai portoenterostomy (KP) can restore bile drainage, although its success decreases with increasing age at surgery.

Case Presentation: We report two infants who underwent intraoperative cholangiography followed by KP due to suspected BA. Liver biopsies confirmed the diagnosis. Outcomes were influenced by the timing of surgery: one patient, who underwent surgery at a later age, experienced an unfavorable course and required early liver transplantation.

Conclusion: These cases highlight age at KP as a key determinant of BA outcomes. Late diagnosis remains a challenge despite existing policies at various levels of care, emphasizing the need to strengthen national screening programs.

Keywords: Biliary Atresia; Hepatic Portoenterostomy; Liver Transplantation; Case Reports (Source MeSH)

Cite as:
Fiestas K, Geraige W, Torres M, Roitman P, Arco K, Vaquer G. Biliary atresia: variability in evolution after Kasai portoenterostomy. Report of two cases. Investig Innov Clin Quir Pediatr. 2025;3(2):37-41.
doi: 10.59594/iicqp.2025.v3n2.145

Corresponding author:
Kerly Fiestas
Telephone: +542614609362
Email: kerlymfiestas@gmail.com

ORCID iDs

Kerly Fiestas
https://orcid.org/0009-0002-6128-5647
Wilma Geraige
https://orcid.org/0009-0009-1652-1947
Mariana Torres
https://orcid.org/0009-0006-8086-7457
Paula Roitman
https://orcid.org/0009-0002-3471-5170
Karina Arco
https://orcid.org/0009-0002-4964-6577

Received : 06/09/2025
Accepted : 09/17/2025
Published : 11/05/2025

Atresia de vías biliares: variabilidad en su evolución posterior a portoenterostomía de Kasai. Reporte de dos casos

RESUMEN

Antecedentes: La atresia de vías biliares (AVB) es una de las causas más frecuentes de colestasis neonatal y la principal indicación de trasplante hepático en niños. Su etiología es desconocida y se caracteriza por la obliteración de los conductos biliares, con interrupción del flujo que se manifiesta como ictericia, coluria, acolia, hepatomegalia y/o esplenomegalia. La portoenterostomía de Kasai (PK) puede restablecer el drenaje biliar, aunque su eficacia disminuye con la edad del paciente.

Descripción de los casos: Se presentan los casos de dos lactantes en quienes, ante la sospecha de AVB, se realizó colangiografía intraoperatoria seguida de PK. Las biopsias hepáticas confirmaron el diagnóstico. La evolución estuvo determinada por el momento quirúrgico: uno de los pacientes, intervenido tardíamente, presentó evolución desfavorable y requirió trasplante hepático precoz.

Conclusión: Estos casos resaltan la edad al momento de la PK como factor determinante en la evolución de la AVB. La detección tardía continúa siendo un desafío, a pesar de las políticas vigentes en los distintos niveles de atención, lo que subraya la necesidad de fortalecer los programas nacionales de cribado.

Palabras clave: Atresia Biliar; Portoenterostomía Hepática; Trasplante de Hígado; Informe de caso (Fuente: DeCS)MeSH)

INTRODUCCIÓN

Biliary atresia (BA) is a severe liver disease of uncertain etiology, resulting from progressive fibrosclerotic obliteration of the intrahepatic and extrahepatic bile ducts. This process leads to progressive jaundice, dark urine, and acholic stools between the second and sixth week of life. Laboratory findings include elevated direct bilirubin and liver enzymes.

Diagnosis requires excluding other causes of neonatal cholestasis to ensure timely recognition and referral. Without intervention, BA progresses to secondary biliary cirrhosis, portal hypertension, and end-stage liver disease, ultimately requiring liver transplantation (LT). (1,2) Its incidence ranges from 0.52 to 1.06 per 100,000 live births and has been reported worldwide, with higher frequency in Asian countries. (3,4)

Its heterogeneous etiology remains under debate. Some lines of evidence suggest associations with perinatal infections, toxic exposures, genetic predisposition, immunological or autoimmune disorders, maternal microchimerism, vascular alterations, and morphogenetic defects. Several clinical phenotypes are currently recognized, including isolated BA, syndromic BA associated with other malformations, cystic BA, and BA associated with cytomegalovirus infection. (5)

Although the absence or contraction of the gallbladder on abdominal ultrasound may suggest the diagnosis, confirmation requires intraoperative cholangiography, followed by Kasai portoenterostomy (KP). This surgical procedure involves resecting the obstructed bile duct and anastomosing a loop of the small intestine to the hepatic hilum to restore bile flow.

Liver biopsy is another key diagnostic tool. Histological findings include ductular proliferation, hepatocellular stasis in small bile ducts, fibrosis, and occasionally giant-cell transformation. The introduction of KP changed the prognosis of the disease, with its effectiveness being highly dependent on the timing of the procedure. (6)

Several adjuvant medical therapies have been implemented, including oral antibiotic prophylaxis, corticosteroids administered for variable durations, and ursodeoxycholic acid after surgery, to stimulate bile flow, prevent cholangitis, and reduce hepatic inflammation and progressive fibrosis. However, while these therapies have reduced bilirubin levels within six months postoperatively, they have not demonstrated improvement in native liver survival.

The success of KP depends not only on the patient's age at surgery but also on factors such as the extent of hepatic damage (fibrosis), presence of cirrhosis, associated anomalies in other organs, surgical team expertise, and dynamic postoperative follow-up.(7,8)

To raise awareness of this rare condition and emphasize its consideration in the differential diagnosis of cholestasis, we present two clinical cases of BA diagnosed in 2023 at Hospital Humberto Notti, Mendoza, Argentina.

CASE 1

Female patient, eutrophic, with a history of neonatal jaundice. At 50 days of life, she was referred to our center due to persistent jaundice associated with dark urine, acholic stools, and hepatomegaly, raising suspicion of BA.

Laboratory tests at diagnosis showed mild anemia, thrombocytosis, and altered liver profile (total bilirubin (TBil): 10.6 mg/dL, direct bilirubin (DBil): 8.8 mg/dL, aspartate aminotransferase (AST): 453 U/L, alanine aminotransferase (ALT): 229 U/L, gamma-glutamyl transferase (GGT): 367 U/L, alkaline phosphatase (ALP): 926 U/L). Liver function assessed through prothrombin time (PT), activated partial thromboplastin time (aPTT), international normalized ratio (INR), and albumin was within normal ranges. Viral serologies were negative, and abdominal ultrasound revealed a contracted gallbladder.

At 55 days of life, diagnostic intraoperative cholangiography was performed, followed by KP. Histopathological findings of the liver biopsy were consistent with BA.

In the immediate postoperative period, the patient developed fever and cholangitis 10 days after surgery, with no pathogen isolated (Table 1). Twenty-four days after surgery, she was discharged with a mild decrease in bilirubin, transaminases, and ALP (TBil: 9.5 mg/dL, DBil: 8.6 mg/dL, AST: 221 U/L, ALT: 177 U/L, ALP: 758 U/L), although GGT showed an increase (3,934 U/L).

During outpatient follow-up one month after KP, the patient demonstrated clinical improvement, characterized by a marked decline in bilirubin levels. At two and a half months, remission of symptoms and normalization of bilirubin levels were observed (Table 2). After a two-year follow-up, she continued to show favorable evolution, receiving antibiotic prophylaxis for cholangitis and treatment with ursodeoxycholic acid. This patient experienced a favorable outcome, with symptom remission following restoration of bile flow through the KP.

CASE 2

A male infant with a history of jaundice since birth, bilateral hydrocele, umbilical hernia, and poor weight gain presented at 2 months of age to the emergency department with obstructive bronchitis. On examination, he was found to have jaundice, dark urine, acholic stools, and hepatosplenomegaly, raising suspicion of BA and prompting hospitalization for further evaluation.

Initial laboratory evaluation showed mild anemia, with TBil at 6.4 mg/dL, DBil at 5.7 mg/dL, AST at 108 U/L, ALT at 38 U/L, GGT at 380 U/L, and ALP at 396 U/L, while liver function remained preserved. Viral serologies were negative, and an abdominal ultrasound indicated the absence of the gallbladder.

After resolution of the acute respiratory illness, at 76 days of life, diagnostic intraoperative cholangiography followed by KP was performed. Histopathological findings of the liver biopsy were consistent with BA. In the immediate postoperative period, he developed fever and cholangitis at 3 weeks, with no organism isolated, requiring a 14-day course of antibiotics (Table 1).

He was discharged 25 days after surgery with TBil at 2.8 mg/dL, DBil at 2.7 mg/dL, AST at 83 U/L, ALT at 51 U/L, ALP at 314 U/L, and GGT at 1,308 U/L. However, his course was

Table 1. Clinical characteristics at diagnosis and evolution of the reported cases

Characteristic	Case 1	Case 2
Reason for consultation	Cholestasis	Respiratory infection
Age at diagnosis (days)	50	60
Nutritional status at diagnosis	Eutrophic	At risk of malnutrition
Associated anomaly	No	Yes
Age at Kasai portoenterostomy (days)	55	76
Restoration of bile flow after KP	Yes	No
Perioperative complications[a]		
Surgical	No	No
Medical (cholangitis)	Yes	Yes
Complications between 2 and 6 months after KP		
Cholangitis (n)	0	2
Portal hypertension	No	Yes
Cirrhosis	No	Yes
APRI index[b] at 5.5 months post-KP	0.4	6.4
PELD score[c] at 6 months post-KP	3	30
Liver transplantation at 6 months post-KP	No	Yes

a. Defined as complications occurring within 30 days after Kasai portoenterostomy (KP).
b. Aspartate Aminotransferase-to-Platelet Ratio Index (APRI). A value <0.5 indicates low probability of significant fibrosis, and >1.5 indicates high probability of advanced fibrosis or cirrhosis.
c. Pediatric End-Stage Liver Disease (PELD) score, used to assess the need for liver transplantation in children under 12 years of age.

unfavorable. Two months after KP, he was readmitted with suspected cholangitis and anastomotic dysfunction, showing persistent symptoms and rising bilirubin and liver enzymes (TBil: 11.2 mg/dL, DBil: 10.3 mg/dL, AST: 160 U/L, ALT: 69 U/L, ALP: 639 U/L, GGT: 235 U/L). Additionally, he developed a urinary tract infection caused by Candida albicans and an upper respiratory infection related to COVID-19 (Table 2).

At 4 months post-KP, he was admitted for the third time in poor overall condition with hepatic failure, interpreted as KP dysfunction. Laboratory tests revealed hematocrit at 27%, hemoglobin at 9.4 g/dL, platelet count of 16,880/mm³, leukocyte count of 12,260/mm³, total protein at 5.6 g/dL, TBil at 14.4 mg/dL, DBil at 13.1 mg/dL, AST at 175 U/L, ALT at 65 U/L, GGT at 110 U/L, and ALP at 580 U/L. An abdominal Doppler ultrasound showed a heterogeneous liver, splenomegaly, ascites, and signs of portal hypertension.

While arrangements were being made for referral to a higher-complexity center for LT, he needed admission to the Intensive Care Unit due to refractory ascitic-edematous syndrome, variceal bleeding, and other infectious and hematologic complications. With a PELD score of 30, INR of 1.6, albumin

Table 2. Biochemical characteristics of the reported cases

Characteristic	Case1	Case 2
Laboratory tests at 30 days post-KP*		
Bilirrubina directa (mg/dL)	3.9	2.9
AST (U/L)	245	92
ALT (U/L)	369	53
GGT (U/L)	3,333	1,000
ALP (U/L)	283	347
Albumin (mg/dL)	3.5	3.6
Laboratory tests at 75 days post-KP*		
Direct bilirubin (mg/dL)	0.7	10.9
AST (U/L)	75	192
ALT (U/L)	54	72
GGT (U/L)	638	126
ALP (U/L)	549	582
Albumin (mg/dL)	3.7	2.7

** Kasai portoenterostomy*
AST: aspartate aminotransferase
ALT: alanine aminotransferase
GGT: gamma-glutamyl transferase
ALP: alkaline phosphatase

at 2.6 g/dL, TBil at 36 mg/dL, and DBil at 33 mg/dL, he was transferred at 7 months and 3 weeks of age to a higher-complexity center, where he underwent LT with a split left-lobe cadaveric donor graft at 8 months of age.

Postoperatively, he experienced several complications, including arterial hypertension, cytomegalovirus reactivation, primary Epstein–Barr virus infection, and an abdominal abscess caused by Klebsiella oxytoca. At 8 months after transplant, he developed a cow's milk protein allergy, associated with patient-specific factors such as the transplant itself, immunosuppressive therapy, organ-specific immune properties, and alterations in the microbiome.

The patient was jointly managed with the transplant center, and after two years of follow-up, remains in good clinical condition under immunosuppressive therapy with tacrolimus.

This patient had an unfavorable course, with no remission of symptoms or restoration of bile flow after KP, requiring LT within the first months of life.

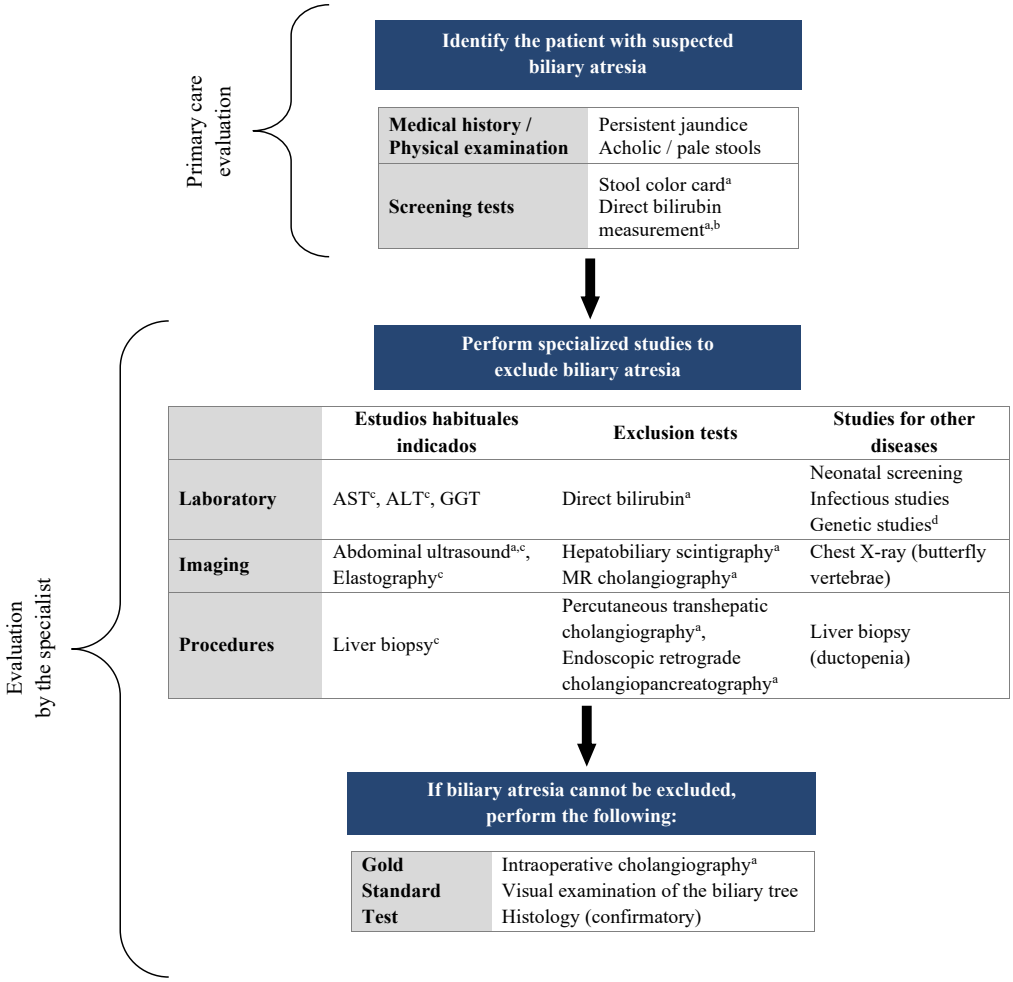
DISCUSSION

BA is a severe disease that manifests within the first three months of life and accounts for 40–50% of all pediatric liver transplants worldwide. It consists of a complete obliteration of the biliary tract with interruption of bile flow, clinically presenting with jaundice, acholia, and hyperbilirubinemia. For diagnosis, a comprehensive assessment of clinical and analytical findings, together with intraoperative cholangiography, is essential, in accordance with the latest clinical practice guidelines (Annex 1). (1,7,9) In the effort not

only to achieve an early diagnosis but also to establish a prognosis after KP, several biomarkers have been investigated. MMP-7 (also known as matrilysin, an enzyme of the matrix metalloproteinase family), IL-33, and GGT have shown utility in facilitating diagnosis. In contrast, the AST-to-platelet ratio index (APRI) can be used to predict significant hepatic fibrosis and post-KP cirrhosis. (10)

Early detection and timely surgical treatment are essential; therefore, age is considered a key prognostic indicator of bile drainage

Appendix 1. Diagnostic algorithm for biliary atresia (adapted from Mysore *et al.*, 2019)



Primary care physicians use clinical signs or screening tests to identify and refer patients with suspected biliary atresia (BA). Specialists perform a series of tests to exclude BA, grouped into three categories: routine tests (a normal result does not rule it out), exclusion tests (a normal result rules it out), and tests for other diseases. If BA cannot be excluded, invasive reference tests are performed. a. Tests that detect biliary abnormalities and may contribute to early diagnosis. b. Screening that parents can also perform. c. Tests that detect liver damage. d. Tests that should be performed promptly. AST: aspartate aminotransferase. ALT: alanine aminotransferase. GGT: gamma-glutamyl transferase.

after KP, as observed in the cases presented. When KP is performed before 2 months of age, there is approximately a 90% probability of restoring bile flow; between 2 and 3 months, the rate is about 40%; and between 3 and 4 months, it is only around 10%. After 4 months of age, the likelihood of restoring bile flow is practically null, which is why the indication of KP in this age group remains controversial. (1,6,11) However, even when KP is performed early, outcomes regarding the need for LT are variable. One third of patients survive more than 10 years without transplantation; another third restore bile flow but require LT before the age of 10; and the remaining third fail to restore bile flow and need transplantation within the first months of life. (12,13)

Adequate and dynamic postoperative follow-up not only significantly prolongs survival but also helps postpone LT and reduce complications such as cholangitis, portal hypertension, hepatopulmonary syndrome, or pulmonary hypertension, as well as prevent, in the long term, the development of hepatic cysts and tumors (hepatocellular carcinoma, hepatoblastoma, and cholangiocarcinoma). (14) Despite the short follow-up period in these two reports and the potential presence of uncontrolled factors, both patients experienced early cholangitis after KP. However, the second case had a greater number of episodes. According to previous findings, this could be related to elevated pre-KP GGT levels, older age at the time of surgery, and more advanced hepatic fibrosis. (15)

These two cases highlight age at the time of KP as a determining factor for subsequent outcomes. However, since only two patients are described, a causal relationship cannot be established, and the findings should not be generalized to other patients. Moreover, this report highlights that delayed detection remains a persistent issue, despite the various policies implemented across different levels of care. For this reason, recent studies have focused on developing strategies to strengthen national screening programs, among which the most notable are the detection of persistent jaundice, determination of fractionated bilirubin in neonates with persistent jaundice at two weeks of life, measurement of bile acid levels on dried blood spot cards, and stool color evaluation. The latter remains the most widely used method worldwide and in our country, where its implementation is crucial for the early detection of cholestasis. (16,17)

Author contributions

All authors participated in the following tasks: Conceptualization, Methodology, Data curation, Formal analysis, Writing – original draft, and Writing – review & editing.

Conflicts of interest

The authors declare no relevant financial or non-financial conflicts of interest.

Funding

This research was self-funded.

Ethical aspects

This case report was approved by the Institutional Research Ethics Committee of the center where the patient was treated.

REFERENCES

1. Mysore KR, Shneider BL, Harpavat S. Biliary atresia as a disease starting in utero: implications for treatment, diagnosis, and pathogenesis. J Pediatr Gastroenterol Nutr. 2019;69(4):396-403. doi:10.1097/MPG.0000000000002450
2. Gorbatyuk O, Kurylo H. Biliary atresia in children (analytical literature review and review of own observation). Wiad Lek. 2024;77(3):577-84. doi:10.36740/WLek202403129
3. Chung PHY, Zheng S, Tam PKH. Biliary atresia: East versus West. Semin Pediatr Surg. 2020;29:150950. doi:10.1016/j.sempedsurg.2020.150950
4. Schreiber RA, Harpavat S, Hulscher JBF, Wildhaber BE. Biliary atresia in 2021: epidemiology, screening and public policy. J Clin Med. 2022;11(4):999. doi:10.3390/jcm11040999
5. Davenport M, Muntean A, Hadzic N. Biliary atresia: clinical phenotypes and aetiological heterogeneity. J Clin Med. 2021;10(23):5675. doi:10.3390/jcm10235675
6. Okubo R, Nio M, Sasaki H, Japanese Biliary Atresia Society. Impacts of early Kasai portoenterostomy on short-term and long-term outcomes of biliary atresia. Hepatol Commun. 2021;5:234-43. doi:10.1002/hep4.1615
7. Ando H, Inomata Y, Iwanaka T, et al. Clinical practice guidelines for biliary atresia in Japan: a secondary publication of the abbreviated version translated into English. J Hepatobiliary Pancreat Sci. 2021;28(1):55-61. doi:10.1002/jhbp.816
8. Ramonet M, Ciocca M, Álvarez F. Biliary atresia: a severe disease. Arch Argent Pediatr. 2014;112(6):542-7. doi:10.5546/aap.2014.542
9. Yoeli D, Choudhury RA, Sundaram SS, Mack CL, Roach JP, Karrer FM, et al. Primary vs. salvage liver transplantation for biliary atresia: a retrospective cohort study. J Pediatr Surg. 2022;57:407-13. doi:10.1016/j.jpedsurg.2023.09.035
10. He L, Ip DKM, Tam G, et al. Biomarkers for the diagnosis and post-Kasai portoenterostomy prognosis of biliary atresia: a systematic review and meta-analysis. Sci Rep. 2021;11(1):11692. doi:10.1038/s41598-021-91047-2
11. Antala S, Taylor SA. Biliary atresia in children: update on disease mechanism, therapies and patient outcomes. Clin Liver Dis. 2022;26(3):341-54. doi:10.1016/j.cld.2022.03.001
12. Hukkinen M, Ruuska S, Pihlajoki M, Kyronlahti A, Pakarinen MP. Long-term outcomes of biliary atresia patients surviving with their native livers. Best Pract Res Clin Gastroenterol. 2022;56-7:101764. doi:10.1016/j.bpg.2021.101764
13. Fuchs J, Mrad C, Gonzales E, Ndiaye D, Fouquet V, Héry G, et al. Biliary drainage surgery before or after 3 months of life versus primary liver transplantation in children with biliary atresia: comparative cohort study. BJS Open. 2023;7(2):175. doi:10.1093/bjsopen/zrac175
14. Calinescu AM, Wilde JC, Korff S, McLin VA, Wildhaber BE. Perioperative complications after Kasai hepatoportoenterostomy: data from the Swiss National Biliary Atresia Registry. Eur J Pediatr Surg. 2020;30:364-70. doi:10.1055/s-0039-1692686
15. Liu J, Dong R, Chen G, Dong K, Zheng S. Risk factors and prognostic effects of cholangitis after Kasai procedure in biliary atresia patients: a retrospective clinical study. J Pediatr Surg. 2019;54:2559-64. doi:10.1016/j.jpedsurg.2019.08.026
16. Ramonet M, Gómez S, Morise S, Parga L, Caglio P, De Micheli M, et al. Early detection of neonatal cholestasis by stool color card screening. Arch Argent Pediatr. 2013;111(2):135-9. doi:10.5546/aap.2013.eng.135
17. Rabbani T, Guthery SL, Himes R, Shneider BL, Harpavat S. Newborn screening for biliary atresia: a review of current methods. Curr Gastroenterol Rep. 2021;23(12):28. doi:10.1007/s11894-021-00825-2