Sri Lanka Journal of Medicine **SLJM**

Case Report

Citation: Prihaningtyas RA, Setyoboedi B, Arief1f S. 2025. Successful Steroid Therapy on Extrahepatic Cholestasis, Sri Lanka Journal of Medicine pp 23-26
DOI: https://doi.org/10.4038/sljm.v34i3.582

Successful Steroid Therapy on Extrahepatic Cholestasis

RA Prihaningtyas[1,2], B Setyoboedi[1,2], S Arief[1,2]

[1]Department of Child Health, Dr. Soetomo General Academic Hospital, Surabaya, Indonesia
[2]Department of Child Health, Faculty of Medicine - UNIVERSITAS AIRLANGGA. Surabava. Indonesia

Correspondence:
B Setyoboedi
E mail:Bagus.setyoboedi@fk.unair.ac.id
https://orcid.org/0000-0002-3923-6913

ABSTRACT

Cholestasis is a pathological jaundice that mostly caused by biliary atresia. Although Kasai surgery is the standard therapy, new developments in adjunctive therapies are still needed to improve outcomes. A 2-month-old baby boy presented with jaundice since 1 week of age, followed by abdominal distention and pale stools. Examination showed hepatomegaly, anaemia, elevated bilirubin, elevated liver enzyme, and reactive Rubella IgG. Liver biopsy indicates extrahepatic cholestasis. The patient received ursodeoxycholic acid and methylprednisolone. After eight weeks, his stool darkened, jaundice resolved, and liver function normalized. This case demonstrates the successful treatment of cholestasis suggesting biliary atresia with steroids.

Keywords: Cholestasis, biliary atresia, jaundice

INTRODUCTION

The progressive inflammatory fibrosclerotic disease known as biliary atresia affects the biliary system[1]. Biliary atresia is a common cause of pathological jaundice in infants. Direct or conjugated bilirubin screening for infants has been showed to result in early diagnosis. Despite the Kasai operation, new developments in adjunctive therapies are still required to improve outcomes (2).

CASE REPORT

A 2-month-old baby boy presented to the hepatology clinic referred from a regional hospital with a chief complaint of jaundice. The jaundice had appeared for 1 week of age. According to the mother, the whole body looked yellow with pale stool colour (Figure 1).

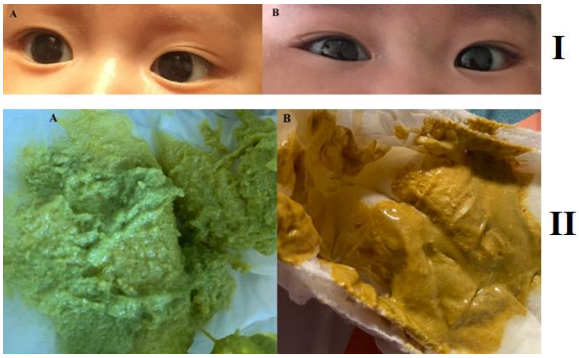

Figure 1. Clinical manifestation of the patient (IA) Before therapy; (IB) After therapy; Improvement of stool colour after therapy (IIA) Before therapy; (IIB) After therapy

Received: 2025-02-01 **Accepted revised version: 2025-10-23** **Published: 2025-11-11**

Other complaints such as enlarged abdomen, bleeding, vomiting, fever was denied. At the age of 1 month and 2 weeks, the patient was brought to a paediatrician's practice, but the doctor said it was physiological jaundice and there was no specific treatment. The patient was born spontaneously, 36 weeks with a birth weight of 2400 grams and a body length of 50 cm.

Physical examination revealed icteric sclera, abdominal distention with hepatomegaly measuring 2x2x1 cm. Laboratory examination showed Hemoglobin 9.7 g/dL, Hematocrite 28.1 %, Leucocyte 9.55 x 10^3/µL, Thrombocyte 408 x 10^3/µL, AST 61 U/L, ALT 33 U/L, Gamma GT 103.0 U/L, Alkaline Phosphatase 922 U/L, total bilirubin 6.3 mg/dL, direct bilirubin 3 mg/dL, Rubella IgG Reactive (2.34 IU/mL), Rubella IgM non-reactive (1.01 AU/mL), CMV IgG Reactive (3.66 AU/mL), CMV IgM non-reactive (0.321 AU/mL), Toxoplasma IgG non-reactive (0.018 IU/mL), and Toxoplasma IgM non-reactive (0.904 AU/mL).

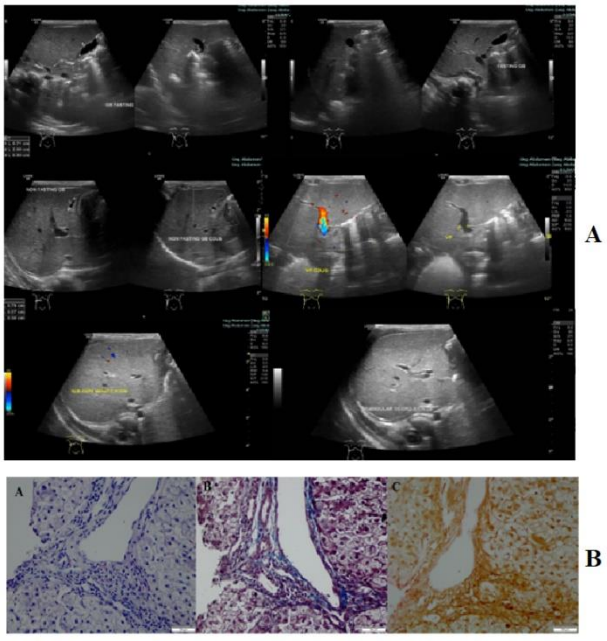

Figure 2. A: A two-phase abdominal ultrasound of the patient; B. Liver histopathological Examination (A) HE 400x; (B) MT 400x; (C) RC 400x

On 2-phase abdominal ultrasound, there was no gallbladder (optimally fasted as recommended). Sonography showed ghosting gallbladder with increased HA/PV ratio (Figure 2A). Liver biopsy examination showed hepatic biopsy tissue pieces consisting of 7 portal tracts. The portal tracts showed proliferation of partially dilated bile ducts containing bile pigment accompanied by a small infiltrate of lymphocyte cells limited to the portal tract. The hepatic lobules were composed of hepatocytes with ballooning degeneration. MT/RC staining showed mild fibrosis in the portal tract (F1) (Figure 2B).

Intraoperative cholangiography was not performed and the patient's parents refused Kasai's operation due to financial constraints. The patient was treated solely with methylprednisolone and ursodeoxycholic acid orally. After administration of steroids and ursodeoxycholic acid for 8 weeks, there was a decrease in liver function test to normal levels (Figure 3). Clinically, the patient was jaundice-free and there was an improvement in stool colour to darker (Figure 1).

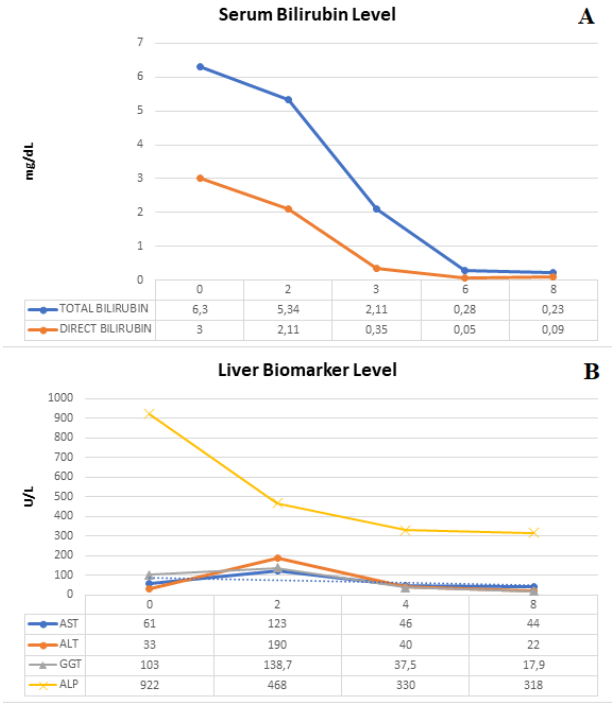

Figure 3. A: Bilirubin serum level improvement after therapy (weeks); B: Liver function test improvement after therapy (weeks)

DISCUSSION

Infants developed with biliary atresia have obliterated intrahepatic and extrahepatic bile ducts, which can cause cholestasis and, if left untreated, cirrhosis. Cholestasis is a manifestation of hepatobiliary dysfunction and is always

pathological (3). Total serum bilirubin and direct bilirubin levels should be measured in order to assess for cholestasis in any infant who develops jaundice after two weeks of age. Elevated serum direct bilirubin levels (>1.0 mg/dL or >17 µmol/L) are indicating of cholestasis (3). The patient's serum bilirubin level was measured and found to be elevated (total bilirubin 6.3 mg/dL, direct bilirubin 3 mg/dL), supporting cholestasis (Figure 3A).

Serum gamma-glutamyl transpeptidase levels were higher in biliary atresia (542 ± 130 vs. 139 ± 25.8 U/L in non-biliary atresia, P = 0.03) (4). According to studies, normal serum GGT levels (12.7%) can occur in biliary atresia. As in this case, the GGT level was found to be < 250 U/L (Figure 3B). Atrophic or hypoplastic gallbladder (GB) is the most common ultrasonographic finding in biliary atresia (65%), but the gallbladder appears normal in 30% of cases (5). With 92% sensitivity and 73% specificity, the absence of the gallbladder on abdominal ultrasonography is a reliable predictor of gallbladder absence during surgery (6). The most sensitive gallbladder abnormality was the absence of contraction after feeding (89%), which was followed by wall abnormality (83%), absence or length <1.5 cm (79%), and gallbladder absence (28%) (6). In this case, a 2-phase abdominal ultrasound showing ghosting gallbladder with increased HA/PV ratio supporting the feature of biliary atresia (Figure 2A).

Intraoperative cholangiography is necessary to diagnose biliary atresia in infants who present with cholestatic jaundice. However, not all health centres are able to perform intraoperative cholangiography routinely. When there is a high suspicion of biliary atresia, liver biopsy remains the mainstay of the diagnostic process. Liver biopsy has a diagnostic accuracy rate of 86–95% for biliary atresia when examined by qualified pathologists (7,8). Bile duct proliferation (92%), canalicular cholestasis (96%), bile plugs (84%), and portal fibrosis (95%), were the most frequently observed histopathologic findings (5). As in this case, liver biopsy showed proliferation of partially dilated bile ducts containing bile pigment accompanied by a small infiltration of lymphocyte cells confined to the portal tract in support of extrahepatic cholestasis with mild fibrosis in the portal tract (F1) (Figure 2B).

Delayed referral for cholestasis is still high in Indonesia. There are still many cases of infants who develop jaundice at 1 month of age but visit the referral hospital at >4 months of age (9). For the purpose of resolving biliary obstruction and improving native liver survival, early diagnosis and Kasai surgery are essential (10). However, studies suggest that only 50-75% of patients can have their bile flow restored with timely Kasai surgery (1). The immune system is thought to be involved in the process of bile duct obstruction in biliary atresia. The randomised control trial study proved that there was an improvement in bilirubin and inflammatory marker after methylprednisolone compared to the placebo group (11). Further researches is needed to determine the effectiveness of steroids in preventing bile duct obstruction and further liver damage due to biliary atresia, especially in limited health facilities that are limited in performing Kasai surgery and liver transplantation.

Conclusion

To prevent more liver damage, based on the immune-mediated mechanism of biliary atresia disease, there is hope that additional therapies such as steroid to suppress inflammation in the bile ducts will be necessary to suppress bile duct obstruction and further liver damage.

Author declaration

Acknowledgement
None

Authors' contributions:
Planning, Gathering Data, Analyzing, Writing: RAP, BS, SA.

Conflicts of interest:
The authors declare that there is no financial or non-financial conflict of interest.

Funding statement:
Self-funded.

Ethics statement:
Written informed consent was obtained from the patient before they participated in the case, ensuring their understanding of the purpose, procedures, and potential implications involved.

Statement on data availability:
All data generated during the study are included

REFERENCES

1. Tam PKH, Wells RG, Tang CSM, Lui VCH, Hukkinen M, Luque CD, et al. Biliary atresia. Nat Rev Dis Primer. 2024 Jul 11;10(1):47. DOI: 10.1038/s41572-024-00533-x
2. Yerina SE, Ekong UD. Biliary Atresia/Neonatal Cholestasis. Pediatr Clin North Am. 2021 Dec;68(6):1333–41. DOI: 10.1016/j.pcl.2021.08.002
3. Fawaz R, Baumann U, Ekong U, Fischler B, Hadzic N, Mack CL, et al. Guideline for the Evaluation of Cholestatic Jaundice in Infants: Joint Recommendations of the North American Society for Pediatric Gastroenterology, Hepatology, and Nutrition and the European Society for Pediatric Gastroenterology, Hepatology, and Nutrition. J Pediatr Gastroenterol Nutr. 2017 Jan;64(1):154–68. DOI: 10.1097/MPG.0000000000001334
4. Robie DK, Overfelt SR, Xie L. Differentiating biliary atresia from other causes of cholestatic jaundice. Am Surg. 2014 Sep;80(9):827–31.
5. Alhebbi H, El-Edreesi M, Abanemai M, Saadah O, Alhatlani M, Halabi H, et al. Clinical and laboratory features of biliary atresia and patterns of management practices: Saudi national study (2000–2018). Saudi J Gastroenterol. 2024 Mar;30(2):89–95. DOI: 10.4103/sjg.sjg_151_23
6. Ho A, Sacks MA, Sapra A, Khan FA. The Utility of Gallbladder Absence on Ultrasound for Children With Biliary Atresia. Front Pediatr. 2021;9:685268. DOI: 10.3389/fped.2021.685268
7. Ahmed ABM, Fagih MA, Bashir MS, Al-Hussaini AA. Role of percutaneous liver biopsy in infantile cholestasis: cohort from Arabs. BMC Gastroenterol. 2021 Dec;21(1):118. DOI: 10.1186/s12876-021-01699-4
8. Russo P, Magee J, Anders R, Bove K, Chung C, Cummings O, et al. Key histopathological features of liver biopsies that distinguish biliary atresia from other causes of infantile cholestasis and their correlation with outcome: a multicenter study. Am J Surg Pathol. 2016 Dec;40(12):1601–15. DOI: 10.1097/PAS.0000000000000755
9. Setyoboedi B, Prihaningtyas RA, Winahyu AK, Arief S. Delayed Admission in Neonatal Cholestasis. J Community Med Public Health Res. 2024 Nov 12;5(2):121–7. **https://doi.org/10.20473/jcmphr.v5i2.47315**
10. Patel KR. Biliary atresia and its mimics. Diagn Histopathol. 2023 Jan;29(1):52–66. DOI: 10.1016/j.mpdhp.2022.11.001
11. Setyoboedi B, Utomo MT, Prihaningtyas RA, Arief S. Effectiveness of oral methylprednisolone as adjuvant therapy for clinical improvement, biochemical markers, and inflammation in infants with cholestasis. Heliyon. 2024 Jul;10(14):e34110. DOI: 10.1016/j.heliyon.2024.e34110