



ORIGINAL RESEARCH PAPER

Paediatrics

EARLY DIAGNOSIS AND SURGICAL MANAGEMENT OF BILIARY ATRESIA IN A NEONATE PRESENTING WITH PERSISTENT CHOLESTATIC JAUNDICE

KEY WORDS: Biliary Atresia, Neonatal Cholestasis, Kasai Portoenterostomy, Direct Hyperbilirubinemia, Hida Scan

Dr Nandhini G

Department of Paediatrics, [Rajarajeshwari Medical College and Hospital], [Bengaluru], [India]

Dr Kartik

Department of Paediatrics, [Rajarajeshwari Medical College and Hospital], [Bengaluru], [India]

ABSTRACT

Biliary atresia is a progressive fibro-obliterative disorder of the extrahepatic biliary tree and a leading cause of neonatal cholestasis. Early diagnosis is essential for timely surgical intervention and improved outcomes. We report a 13-day-old term male neonate presenting with persistent jaundice since day 2 of life. Initial evaluation suggested unconjugated hyperbilirubinemia; however, subsequent investigations revealed conjugated hyperbilirubinemia with elevated gamma-glutamyl transferase levels. Imaging studies, including ultrasonography and hepatobiliary iminodiacetic acid (HIDA) scan, demonstrated findings suggestive of extrahepatic biliary obstruction. The diagnosis was confirmed intraoperatively, and the patient underwent Kasai portoenterostomy. This case highlights the importance of early recognition and prompt evaluation of neonatal cholestasis to facilitate timely surgical management and improve prognosis.

BACKGROUND

Biliary atresia is a progressive cholangiopathy characterized by obliteration of the extrahepatic bile ducts, leading to cholestasis, fibrosis, and eventual cirrhosis. It is the most common indication for liver transplantation in children. The incidence varies globally, with higher prevalence reported in Asian populations. Early diagnosis is critical, as surgical intervention with Kasai portoenterostomy performed before 60 days of life significantly improves bile flow and long-term outcomes. Delay in diagnosis results in progressive liver damage and poor prognosis.

Case Presentation

A 13-day-old male neonate, born at term (39+4 weeks) via emergency lower segment cesarean section to a primigravida mother, presented with yellowish discoloration of skin and sclera since 48 hours of life. The baby cried immediately after birth and had a birth weight of 3.42 kg. In the early neonatal period, the baby developed respiratory distress and was managed as transient tachypnea of the newborn with oxygen support, CPAP, and HFNC. Initial bilirubin at 48 hours was 8.4 mg/dL (direct 1.3 mg/dL), for which phototherapy was administered.

On day 9 of life, the jaundice worsened. Investigations revealed total bilirubin of 21.3 mg/dL with direct bilirubin of 4.6 mg/dL. Liver enzymes were mildly elevated, and CRP was negative. Initial ultrasonography was reported as normal. The baby was started on ursodeoxycholic acid and fat-soluble vitamins.

Due to persistent conjugated hyperbilirubinemia, the neonate was referred for further evaluation. On examination, hepatomegaly (2 cm below the costal margin) and a palpable spleen tip were noted. Repeat investigations showed total bilirubin of 21.5 mg/dL with direct bilirubin of 6.2 mg/dL.

Investigations

Further evaluation revealed elevated gamma-glutamyl transferase (GGT) levels (551 IU/L). Repeat ultrasonography demonstrated non-visualization of the common bile duct and a small, irregular gallbladder.

A hepatobiliary iminodiacetic acid (HIDA) scan showed hepatomegaly with impaired tracer uptake and absence of biliary excretion into the intestine up to 24 hours, suggestive of extrahepatic biliary obstruction.

Intraoperative cholangiogram confirmed biliary atresia.

Differential Diagnosis

The following conditions were considered:

- Neonatal hepatitis
- Choledochal cyst
- Progressive familial intrahepatic cholestasis
- Metabolic liver disorders

The presence of conjugated hyperbilirubinemia, elevated GGT, and characteristic imaging findings supported the diagnosis of biliary atresia.

Treatment

The patient underwent Kasai portoenterostomy following confirmation of diagnosis via intraoperative cholangiogram. A liver biopsy was also obtained for histopathological evaluation.

Supportive management included nutritional optimization and supplementation with fat-soluble vitamins.

Outcome and Follow-up

The patient was monitored postoperatively for improvement in bile flow and reduction in bilirubin levels. Long-term follow-up was planned to assess liver function, growth parameters, and the need for potential liver transplantation if disease progression occurs.

DISCUSSION

Biliary atresia is a critical cause of neonatal cholestasis and requires early identification for optimal outcomes. The presence of conjugated hyperbilirubinemia beyond two weeks of life should prompt evaluation for cholestatic disorders.

Elevated GGT levels and imaging findings such as absent bile duct visualization are key indicators. HIDA scan plays a supportive diagnostic role, while intraoperative cholangiogram remains the gold standard.

Kasai portoenterostomy is the primary surgical treatment, with success closely linked to early timing. Delayed intervention leads to progressive fibrosis, cirrhosis, and eventual need for liver transplantation.

This case underscores the importance of early suspicion and structured evaluation of persistent neonatal jaundice.

Patient Consent for Publication

Informed consent was obtained from the patient's parents for publication of this case report.

Competing Interests

None declared.

Contributors

[Dr Nandhini G]: conception, data collection, drafting of manuscript. [Dr kartik]: critical revision of manuscript. All authors approved the final version.

Ethics Statement

This case report was prepared in accordance with standard CARE guidelines. Patient data have been anonymised. Institutional ethics approval was not required for a single case report as per local policy.



Fig 1: Intraoperative Photograph Showing Fibrotic Biliary Remnant at the Porta Hepatitis During Kasai Procedure

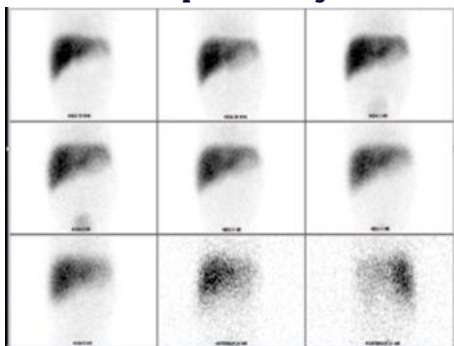


Fig 2: HIDA Scan (Hepatobiliary Iminodiacetic Acid Scan)

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