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Benign Recurrent Intrahepatic Cholestasis (BRIC) Case Series

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ABSTRACT

Background: Benign recurrent intrahepatic cholestasis (BRIC) is a rare autosomal recessive hepatic disorder characterized by intermittent episodes of cholestatic jaundice, pruritus, and fatigue. Despite alarming biochemical derangements, BRIC does not lead to cirrhosis or chronic liver failure. Due to its self-limiting nature and rarity, it is often underdiagnosed, particularly in developing countries.

Aims: To describe the clinical presentation, laboratory features, imaging, histopathological findings, and therapeutic outcomes in five histologically confirmed cases of BRIC and raise awareness of this underrecognized entity.

Methods:A retrospective case series was conducted at a tertiary care centre in Belagavi, India. Five patients diagnosed with BRIC between 2021 to 2024 were studied. Data were obtained from the Department of Gastroenterology and Medical Records, including demographics, symptoms, liver function tests, imaging (USG and MRCP), histopathology, treatment, and outcomes.

Results:All five patients (4 males, 1 female; age range: 14–24 years) presented with recurrent episodes of jaundice and pruritus. Liver function tests consistently showed cholestatic patterns with elevated direct bilirubin and alkaline phosphatase, and relatively preserved transaminases. Viral, autoimmune, and metabolic causes were excluded. Imaging (USG and MRCP) revealed no biliary obstruction or architectural liver damage. Liver biopsy demonstrated canalicular and intrahepatic cholestasis. All patients responded well to supportive management, including ursodeoxycholic acid and cholestyramine, with resolution of symptoms.

Discussion:BRIC, while benign, can cause significant distress due to recurrent jaundice and itching. The episodic nature, lack of imaging findings, and histology-driven diagnosis necessitate a high index of suspicion. In our series, patients responded favourably to conservative management without long-term complications or disease progression.

Conclusion:BRIC should be considered in young patients presenting with episodic cholestatic jaundice and normal imaging findings. Prompt diagnosis through histopathological confirmation avoids unnecessary interventions. Long-term outcomes are excellent with appropriate symptomatic management and patient counselling

Keywords: Benign Recurrent Intrahepatic Cholestasis (BRIC), Cholestatic jaundice, Recurrent jaundice, Ursodeoxycholic acid, Pruritus, Liver biopsy, Intrahepatic cholestasis, ATP8B1 mutation, Canalicular cholestasis, non-cirrhotic liver disease

1. INTRODUCTION

BRIC is a rare but important hepatic condition under the broader spectrum of intrahepatic cholestatic diseases(1). First described by Summerskill and Walshe in 1959, BRIC is now classified into two types: BRIC1, associated with mutations in the ATP8B1 gene, and BRIC2, related to ABCB11 gene mutations(2). Both mutations lead to defective bile acid secretion, causing accumulation of bile within hepatocytes. BRIC is distinguished by its episodic nature, with symptom-free intervals, and absence of progressive fibrosis or cirrhosis(3)

The clinical presentation includes recurrent jaundice, pruritus, fatigue, and anorexia. During episodes, liver function tests reveal a cholestatic pattern—markedly elevated bilirubin and alkaline phosphatase (ALP) with relatively normal or mildly elevated transaminases. Imaging is usually non-contributory(4). Liver biopsy is crucial for diagnosis, showing intrahepatic and canalicular cholestasis. Despite its benign course, BRIC significantly affects quality of life due to debilitating pruritus and cosmetic concern from recurrent jaundice(5).

In India, BRIC remains underreported due to lack of awareness and limited access to genetic testing. This series details five histologically confirmed cases of BRIC from a tertiary care centre in Karnataka, India.

2. MATERIALS AND METHODS

This retrospective study was conducted at the Department of Gastroenterology, KLES Dr. Prabhakar Kore Hospital, Belagavi. Case records of five patients diagnosed with BRIC between 2021 and 2024 were reviewed. Data were obtained from the medical records department and departmental database.

Inclusion criteria were:

Age <30 years

Recurrent jaundice with cholestatic pattern

Normal viral hepatitis and autoimmune profiles

Normal biliary imaging (USG/MRCP)

Histological confirmation of intrahepatic canalicular cholestasis

No evidence of chronic liver disease or cirrhosis

Data collected included demographic details, symptoms, number and duration of episodes, family history, liver function tests, imaging findings, liver biopsy results, and treatment outcomes.

3. CASE PRESENTATIONS

Case 1: 20/M

This young male presented with generalized pruritus, fatigue, anorexia, and jaundice for 1 month. He reported similar self-limiting episodes at 6-month intervals since age 17. On examination, he had deep icterus and excoriation marks, but no hepatosplenomegaly or signs of chronic liver disease. Labs showed TB: 25 mg/dL, DB: 21 mg/dL, ALP: 487 IU/L, SGOT/SGPT mildly elevated. Viral markers were negative. USG and MRCP revealed no biliary obstruction. Liver biopsy revealed canalicular cholestasis without fibrosis. Treated with Ursodeoxycholic acid (UDCA), Solubid (methylprednisolone), and symptomatic. Responded within 7 days; LFT normalized in 4 weeks.

Case 2: 20/M

Presented with third recurrence of jaundice in 2 years. LFT showed TB: 21 mg/dL, DB: 17.2 mg/dL, ALP: 259 IU/L. Imaging showed mild splenomegaly; MRCP was normal. Liver biopsy revealed bland cholestasis. Treated with Heptal, Solubid, and Ursodiol. Discharged after symptomatic resolution. Follow-up showed no evidence of fibrosis or liver failure.

Case 3: 14/M

Admitted with severe pruritus, anorexia, and jaundice. Labs revealed TB: 8.9 mg/dL, ALP: 783 IU/L. Biopsy showed mild ballooning with canalicular bile stasis. Treated with Udivil (UDCA), Choltran (cholestyramine), and antihistamines. Recovered within 2 weeks. Remained symptom-free during 6-month follow-up.

Case 4: 23/F

Presented with right upper quadrant discomfort, pruritus, and jaundice. MRCP ruled out extrahepatic pathology. LFT showed TB: 3.49 mg/dL, ALP: 114 IU/L. Liver biopsy confirmed diagnosis. Managed conservatively with UDCA, Domperidone, Cobadex, and lactulose. Responded well. No recurrence during next 8 months.

Case 5: 23/M

Reported with previous two similar episodes over 4 years. LFTs showed cholestatic pattern; imaging normal. Biopsy revealed bile plugs within dilated canaliculi. Treated with nutritional support and UDCA. Experienced rapid relief from pruritus and jaundice.

Biochemical Profile Summary

Table 1: Liver Function Test Parameters of Selected Patients

Cases	TB (mg/dL)	DB (mg/dL)	SGOT (IU/L)	SGPT (IU/L)	ALP (IU/L)
Case 1 - 20/M	25	21	55	61	487
Case 2 - 20/M	21	17.2	26	18	259
Case 3 - 14/M	8.9	8.3	32	21	783
Case 4 - 23/F	3.49	3.40	53	22	114
Case 5 - 23/M	15	12	20	53	104

4. INVESTIGATIONS

All five patients underwent a standardized evaluation to determine the cause of recurrent jaundice:

Liver Function Tests (LFTs): Revealed a cholestatic pattern with elevated total and direct bilirubin, high alkaline phosphatase (ALP), and normal to mildly raised AST/ALT. Synthetic functions (albumin, INR) were preserved.

Viral & Autoimmune Workup: Negative serologies for hepatitis A, B, C, and E. ANA, AMA, and ASMA were all negative, ruling out viral and autoimmune hepatitis.

Metabolic Screening: Normal serum ceruloplasmin and 24-hour urinary copper excluded Wilson's disease. No findings suggested other metabolic liver diseases.

Imaging:

Ultrasound: Normal liver echotexture, no biliary dilatation; two cases had incidental gallstones.

MRCP: Normal intrahepatic and extrahepatic biliary ducts in all patients.

Liver Biopsy: Showed intrahepatic and canalicular cholestasis without fibrosis or inflammation, confirming BRIC after exclusion of other causes.

5. MANAGEMENT

Treatment focused on symptom relief and cholestasis control:

Ursodeoxycholic Acid (UDCA): Given to all patients; improved bile flow and reduced bilirubin levels.

Cholestyramine: Provided effective relief from pruritus in most patients.

Steroids: Used in selected cases with persistent symptoms.

Supportive Care: Included antihistamines, antiemetics, and nutritional supplements (fat-soluble vitamins).

Follow-Up: All patients improved clinically and biochemically, with no progression to chronic liver disease. Counselling was provided regarding recurrence and the benign nature of BRIC.

6. DISCUSSION

Benign Recurrent Intrahepatic Cholestasis (BRIC) is a rare, autosomal recessive liver disorder characterized by intermittent episodes of cholestasis, typically presenting in adolescence or early adulthood(6). The disorder belongs to the spectrum of familial intrahepatic cholestatic diseases but is differentiated from Progressive Familial Intrahepatic Cholestasis (PFIC) by its non-progressive nature and absence of hepatic fibrosis or liver failure over time.

The pathogenesis of BRIC is linked to mutations in either the **ATP8B1** gene (BRIC type 1) or the **ABCB11** gene (BRIC type 2), which encode proteins involved in bile acid transport. Defects in these genes lead to impaired bile secretion, accumulation of bile salts in hepatocytes, and consequent intrahepatic cholestasis. Despite these disruptions, the hepatocellular architecture remains preserved, distinguishing BRIC histologically and prognostically from more severe cholestatic liver diseases.

Clinically, BRIC presents with recurrent jaundice, severe pruritus, fatigue, and anorexia. Episodes can last from days to weeks and resolve spontaneously, often triggered by infections, hormonal changes, or stress. In our series, all patients had multiple such episodes, and notably, none showed signs of chronic liver disease, hepatosplenomegaly, or portal hypertension. This aligns with the literature, where BRIC is considered a self-limiting but quality-of-life-impacting condition.

The diagnostic approach to BRIC is primarily **exclusion-based**. Common differentials include viral hepatitis, autoimmune hepatitis, drug-induced liver injury, PFIC, Wilson's disease, and biliary obstruction. In our patients, all such causes were systematically ruled out through laboratory panels and imaging. Importantly, **liver biopsy** played a pivotal role, showing classic features of bland intrahepatic and canalicular cholestasis, without inflammation or fibrosis, confirming the diagnosis.

Management is largely symptomatic. **Ursodeoxycholic acid (UDCA)** remains the cornerstone of therapy, improving bile flow and reducing serum bilirubin. **Cholestyramine** was effective in controlling pruritus, and antihistamines provided additional symptom relief. Steroids were reserved for those with persistent symptoms and showed good response. Nutritional support, particularly supplementation of fat-soluble vitamins, is essential during prolonged episodes to prevent deficiencies.

While BRIC is benign and non-progressive, its recurrent nature can cause significant distress and anxiety for both patients and families. This highlights the importance of **patient education and genetic counselling**, especially in populations where consanguinity is prevalent. Although genetic testing was not performed due to resource constraints, the clinical and histopathological profile was sufficient for diagnosis in all cases.

This case series reinforces the importance of **recognizing BRIC early** in the diagnostic pathway of recurrent jaundice in young patients. Awareness among clinicians can avoid extensive and expensive investigations and reassure patients about the excellent long-term prognosis of this condition.

CONCLUSION

BRIC is a benign hepatic condition characterized by episodic intrahepatic cholestasis, commonly affecting adolescents and young adults. It should be considered in the differential diagnosis of recurrent jaundice, particularly when imaging is normal, and hepatitis markers are negative. Liver biopsy is critical for confirmation. Management remains supportive with excellent long-term prognosis. Educating patients about recurrence and benign nature is crucial to improve quality of life and adherence to follow-up.

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