

# Extra-Hepatic portal venous obstruction with portal biliopathy in infant presenting as neonatal cholestasis: a rare case report and review of literature

Yadav A.K.<sup>1</sup>, Meena R.<sup>2</sup>, Bhavika Y.M.<sup>3</sup>, Guruyayappa R.<sup>4</sup>, Dewan V.<sup>5</sup>

<sup>1</sup>Dr Ajeet Kumar Yadav, <sup>2</sup>Dr Ramkesh Meena, <sup>3</sup>Dr Bhavika YM, <sup>4</sup>Dr Guruayappa Ramsingh, <sup>5</sup>Dr Vivek Dewan, all authors are affiliated with Department of Pediatrics, Dr. RML Hospital, New Delhi, India

**Address for Correspondence:** Dr. Ajeet Kumar Yadav, Department of Pediatrics, Dr. RML Hospital, New Delhi, India, Email: ajeet17llrm@gmail.com

## Abstract

Extra-hepatic portal venous obstruction is most common cause of portal hypertension in children. Apart from variceal formation in gastric and esophageal mucosal layer, it has been postulated that external pressure and or ischemic injury play a role in portal biliopathy. We report an unusual case of a 6 month old infant who presented to us with clinical features suggestive of extra-hepatic biliary atresia; diagnosed as EHPVO with portal biliopathy, managed surgically.

**Keywords:** Extra-hepatic portal obstruction, Jaundice, Portal biliopathy

## Introduction

Extra-hepatic portal venous obstruction (EHPVO) is a blockage to the flow of blood in portal vein before it reaches to the liver. EHPVO is the commonest cause of portal hypertension in children which has commonest presentation of upper gastrointestinal bleed [1-3]. There are strictures and dilatations in entire biliary tract and gall bladder secondary to ischemic and compressive effects of portal cavernoma formed in setting of portal

hypertension which presents as clinical features of obstructive jaundice, pruritis, pain abdomen (Portal biliopathy) [4,5]. Several studies have reported that 81-100% of cases of EHPVO have portal biliopathy (PTB) but only one fifth of them present as jaundice [4,6,6-10]. We report a case EHPVO with PTB in a 6 month infant which has never been reported in literature [4,11-13].

## Case Report

A 6 months, female presented to our hospital with yellowish discoloration of eyes and urine since day 6<sup>th</sup> of life along with failure to gain weight, progressive abdominal distension and irritability of 3 month's duration. Jaundice had been deepening since onset. There was history of passage of clay colored stool, which was non bulky and foul smelling since 2<sup>nd</sup> week of life. Abdominal distension was progressive along with poor weight gain. There was no history of hematemesis, melena, itching; antenatal history was uneventful and there was no history of umbilical sepsis or NICU hospitalization. Infant was exclusively breast fed till 2 months of age. There was no history of consanguinity, abortion, stillbirth or similar illness in family. Examination revealed, no dysmorphic facies, with normal fundus, dark yellow sclera and skin with poor muscle bulk without pedal edema or bleeding manifestation. Abdominal examination showed distended abdomen with fullness over right and left hypochondrium. Liver was 6 cm below right costal margin with span of 9 cm and firm in consistency. Spleen was 4 cm below left costal margin. Free fluid was present and bilateral kidneys were not palpable. Cardiovascular, respiratory and central nervous systems were essentially normal.

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Investigation results were; Hemoglobin- 7.1gm/dl, Total leukocyte count- 13900cell/ mm<sup>3</sup>, Polymorphs-34%, Lymphocytes-58%, Eosinophils-8%, Platelet count- 2 lakh/mm<sup>3</sup>, Total serum bilirubin- 18 mg/dl with Direct component- 17.3 mg/dl, Aspartate transaminase- 288 U/L, Alanine transaminase- 514 U/L, Alkaline phosphatase- 373 U/L, Prothombin Time (PT)- 24.1(13.5), International normalized ratio (INR)- 2.85, Activated partial thromboplastin time (aPTT)- 49.3(28.7), TORCH- Negative, Blood urea- 15 mg/dl, Serum creatinine- 0.2 mg/dl.

Hepatitis B surface antigen (HBsAg), antibody to hepatitis C, A and E were negative. Upper gastrointestinal endoscopy showed two columns of grade II varices. Ultrasound abdomen revealed hepatosplenomegaly, gross ascites, contracted gallbladder, periportal cuffing with normal intrahepatic biliary radicles and portal vein diameter of 4mm. Color Doppler study of splenoportal system showed periportal anechoic structures at porta hepatis. CECT abdomen and CT portovenography showed splenomegaly with dilated tortuous splenic vein and collaterals, findings suggestive of EHPVO. A diagnosis of EHPVO with PTB was made on the basis of Magnetic resonance cholangiopancreatography (MRCP) findings showing tortuous biliary tree with multiple biliary strictures in common hepatic duct, common bile duct and cystic duct. Supportive management was started and urgent referral was made to specialized center for surgical management. Outcome in this case was not known as this patient did not turn-up on follow up.



**Figure-1: Infant with pigmented skin with Heato-splenomegaly**

## Discussion

The clinical presentation in this 6 month old infant with onset of jaundice since 6<sup>th</sup> day of life and acholic stool, suggestive of extra-hepatic biliary atresia was later confirmed to be due to PTB as a result of EHPVO. This kind of presentation is unique and has never been reported in the literature, prompted us to report this case. The relationship between EHPVO and jaundice was first time reported by Gibson et al in 1965 [14]. The most common presentation of EHPVO in pediatric age group is well tolerated variceal bleeding and

splenomegaly [4]. It is often recognized when a child is being evaluated for jaundice, pruritus, acute cholecystitis-like syndrome and ascites [7,15,16].

Aetiology of EHPVO in children has not been well documented however omphalitis, neonatal umbilical sepsis, intra-abdominal infection, post umbilical catheterization have been linked to development of EHPVO in children [17].

Extra-hepatic portal vein obstruction (EHPVO) with portal hypertension leads to abnormality in entire biliary tract including intra-hepatic and extra-hepatic bile ducts, cystic duct and gallbladder changes referred as portal hypertensive biliopathy (PTB) [4,18]. Several studies have shown frequency of biliary tract changes in adult patients with EHPVO [6,9,10,19-22], but very less is known about pediatric age group [18]. Although it has been postulated that dilatation of epicholedochal venous plexus of Saint [23] causes fine irregularities in biliary tract and dilatation of paracholedochal venous plexus of Petren [24] causes extrinsic compression over CBD [25]. Post ischemic biliary stricture [25] and encasement of bile duct in solid tumor like cavernoma [21] are other mechanism involved in PTB although mechanism of ischemia in EHPVO remain obscured [25].

In patients with EHPVO, majority of them (80% to 90%) presents with sudden onset hematemesis [1,2,26]. The patient under this case report presented to us with deepening jaundice, abdominal distension, failure to gain weight and presence pale stools. The clinical presentation in this case was related to chronic cholestasis, following biliary obstruction secondary to compression of biliary tract with enlarged collateral and ischemic injury to the lumen (PTB).

Findings of PTB and EHPVO can be well described by colour Doppler flow imaging, endoscopic retrograde cholangiopancreatography (ERCP), CECT and MRI [27-29]. We diagnosed this case with MRCP and CECT portovenography [30,31].

## Conclusion

This unusual age of presentation in this case emphasizes, high index of suspicion and colour Doppler study are helpful tool for the diagnosis of PTB, and however confirmatory diagnosis is established with ERCP or non-invasively by MRCP.

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