

# EXTRAHEPATIC PORTAL VEIN OBSTRUCTION IN CHILDREN; A SINGLE-CENTER EXPERIENCE OF NORTHERNMOST INDIA. A FALLING TREND FROM A PREVIOUSLY ENDEMIC REGION

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DOI: 10.5281/zenodo.10983474

## Abstract

**Background:** Extrahepatic portal vein obstruction (EHPVO) is a common cause of upper gastrointestinal bleeding (UGIB) in children. It could be secondary to neonatal omphalitis, umbilical vein catheterization (UVC), thrombophilic diseases, or idiopathic. This study is done to look for the clinico-etiological profile of EHPVO in the pediatric population. **Methods:** This is a prospective, observational study conducted in the Department of Pediatrics, Govind Ballabh Pant Hospital, J&K, Srinagar, India from December 2019 to December 2021. All the consecutive patients of EHPVO attending the hospital were enrolled. Endoscopy was performed in all cases except infants and findings were noted. Endoscopic banding was done for the bleeders. Contrast-enhanced computed tomography scan (CECT), Magnetic resonance angiography (MRA) of the abdomen, and thrombophilia profile were done in selected patients. **Results:** A total of 15 patients were included, 53.3% were males and 46.7% were female (ratio 1.1:1). The mean age was  $6.91 \pm 3.7$  years. 08/15 (53.3%) were in the age group of 05 to 10 years. BMI was  $<18.5 \text{ Kg/m}^2$  in 9/15 (60%) patients. Gastrointestinal bleed in the form of hematemesis and/or malena was the most common symptom, present in 10/15 (66.7%) patients. 04/15 (26.7%) patients had a history of umbilical vein catheterization in the neonatal period, and 01 patients had a history of umbilical sepsis at birth. Palpable Spleen was the most common sign seen in 07 patients (46.7%). Ultrasound abdomen with doppler revealed Portal Cavernoma in 15/15 patients (100%), and splenomegaly in 13/15 patients (87%) UGI endoscopy was done in 13/15 patients, 12/13 (92.3%) had esophageal varices. 10/12 (83.3%) patients required banding. 01/15 patients had MRA documented portal biliopathy. **Conclusion:** EHPVO is most common in the 05 to 10 years age group. UGI bleed is the most common presenting symptom. Esophageal varices requiring banding is the most prominent finding in upper GI endoscopy. Most cases were idiopathic in our series.

**Keywords:** EHPVO, GI bleed, Esophageal varices.

## INTRODUCTION

Extra hepatic portal vein obstruction (EHPVO) is a vascular disorder of liver. EHPVO may present with symptomatic portal hypertension, portal biliopathy, growth retardation, hepatic dysfunction and bleeding from varices<sup>1-6</sup>. Portal vein obstruction associated with chronic liver disease or neoplasia does not constitute EHPVO<sup>3</sup>. It is idiopathic in more than 50% of cases<sup>7-9</sup> but could be secondary to neonatal omphalitis, umbilical vein catheterization (UVC)<sup>10</sup> and thrombophilic diseases<sup>3,11</sup>. Ultrasonography abdomen with doppler is the investigation of choice. This study is done to look for the clinico-etiological profile of EHPVO in pediatric population

## METHODS

This is a prospective, observational study conducted in the Department of Pediatrics, Govind Ballabh Pant Hospital, J&K, Srinagar, India from December 2019 to December 2021. Diagnosis of EHPVO was based on non-visualization of main portal vein and presence of portal cavernoma on USG Doppler. Patients with non-cirrhotic portal fibrosis or cirrhosis were excluded from the study. Complete anthropometry of all the patients was taken and plotted on the WHO 2006 and IAP 2015 combined growth charts for boys and girls 0 to 18 years of age. Weight of the cases was measured in light clothing without shoes by standing on a calibrated weighing machine. The height of the cases was measured in standing position by calibrated meter scale. Blood samples were taken for complete blood count, liver and renal function tests and coagulation studies. Endoscopy was performed using video endoscope (Olympus N30 upper gastrointestinal fibre optiscope) in all cases except infants to look for esophageal varices, gastric varices and portal hypertensive gastropathy (PHG). Endoscopic banding was done for the bleeders. Contrast enhanced computed tomography scan (CECT) of abdomen was done in 01 patient for suspected pancreatitis. Patients of EHPVO with right upper quadrant pain and jaundice were subjected to Magnetic resonance angiography (MRA) to confirm portal biliopathy. Thrombophilia profile including Protein C and S, Antithrombin 3 and factor v Leiden mutation were done in selected patients. This study was approved by the institutional ethics committee.

Statistical method: Numerical data is presented as mean $\pm$ SD. Frequencies are reported as percentages. The statistics is done using SPSS version 25.

## RESULTS

A total of 15 patients were included, 53.3% were males and 46.7% were female (ratio 1.1:1). The mean age was 6.91 $\pm$ 3.7 years. 08/15 (53.3%) were in the age group of 05 to 10 years. The mean weight and mean height were 25.3 $\pm$ 12 Kg, and 121 $\pm$ 26.5 cm respectively. The mean BMI was 16.1  $\pm$ 2.4 Kg/m<sup>2</sup>. The BMI was <18.5 Kg/m<sup>2</sup> in 09/15 (60%) patients. Gastrointestinal bleed in the form of hematemesis and/or malena was the most common symptom, present in 10/15 (66.7%) patients. Yellowish discoloration of eyes and fever was seen in 2 patients (13.3%). 04/15 (26.7%) patients had history of umbilical vein catheterization in the neonatal period, 01 patient had history of umbilical sepsis at birth. The remaining 10 patients (66.7%) had an unremarkable postnatal and neonatal period life. Palpable spleen was the most common sign presented in 07 patients (46.7%). Ultrasound abdomen with Doppler revealed Portal Cavernoma in all 15/15 patients (100%), splenomegaly in 13 patients (87%) and ascites in 01 patient. UGI endoscopy was done in 13/15 patients, 2 patients being neonates were excluded. Esophageal varices were present in 12/13 (92.4%) patients. 10/12 (83.3%) patients required banding. 01/15 patients had MRA documented portal biliopathy.

In our study, out of 15 patients, 1 patient (6.6%) had chronic pancreatitis, USG abdomen showed portal cavernoma with bulky pancreas. Contrast enhanced computed tomography (CECT) abdomen was done to confirm the findings. This patient had persistently deranged coagulogram. The thrombophilia profile of this patient was normal. The genome sequencing showed congenital afibrinogenemia. Out of 15 patients, 01 patient (6.6%) had portal biliopathy confirmed by MRA

## DISCUSSION

EHPVO is the most common cause of noncirrhotic portal hypertension in India<sup>12</sup> and accounts for 40% cases of portal hypertension in children<sup>13</sup>. In our study 60% patients had growth retardation with BMI less than 18.5 Kg/m<sup>2</sup> which was comparable to the previous observations by Sarin et al<sup>12</sup> and Mehrotra et al<sup>14</sup> who reported that 51% and 54.5% patients with EHPVO had growth retardation respectively. In the neonatal period, 04/15 (26.7%) patients had history of umbilical vein catheterization, and 01 patient had history of umbilical sepsis at birth. The remaining 10 patients (66.7%) had an unremarkable postnatal and neonatal period life. Similar findings were reported by Prapun Aanpreung et al<sup>15</sup>. EHPVO was idiopathic in >50% cases in his cohort and 27% had history of umbilical vein catheterization in their neonatal period. Sushanta Kumar Jena et al<sup>16</sup> also found history of neonatal umbilical sepsis in 15.9% cases and history of umbilical vein catheterization in 10.2% cases together comprising 26% of cases. The most common manifestation in our study was UGI bleed in the form of hematemesis and /or melena seen in 66.7% of cases. On UGI endoscopy, 92.3% cases were found to have esophageal varices. Karthikeyan P et al<sup>17</sup> found that recurrent variceal bleed was the most common presentation in 83.3% cases. This is almost similar to our study. In our study 46.7% cases had palpable spleens on examination and 87% cases had splenomegaly on USG abdomen which is less compared to the observations by Sushanta Kumar Jena<sup>16</sup> and Neal Abd EL Hamid et al<sup>8</sup> respectively. In our study, 13% had raised PT/INR, 20% cases had hypersplenism and 6% cases had ascites which is comparable to the observations by Nehal Abd EL Hamid et al<sup>8</sup>, a study done at King's college Hospital London.

Our study has certain limitations. The number of patients is less. This is due to the decrease in incidence of EHPVO due to increasing health education, better access to health care, more pediatricians and effective management of neonatal and childhood illnesses like umbilical sepsis and dehydration. A larger multi-centric study would be required to confirm our findings.

## CONCLUSION

EHPVO is most common in 05 to 10 years age group. UGI bleed is the most common presenting symptom. Esophageal varices requiring banding is the most prominent finding on upper GI endoscopy. Most cases were idiopathic in our series.

**Conflicts of interest:** None

**Funding:** Nil

**Declarations:**

Ethics approval: The study is approved by the institutional ethics committee of Government Medical college and associated hospitals Srinagar Kashmir under Reference no. 180//ETH/GMC/ICM dated 22<sup>th</sup> May 2019.

Consent to participate: Since this is an observational study, the consent to participate has been waived off by the ethics committee of Government Medical college and associated hospitals Srinagar Kashmir

Consent for publication: Not applicable.

Availability of data and materials: Data and material will be available on request.

Competing interests: There are no competing interests.

Funding: The study is not funded.

### Author contributions:

JH, IM, NA, SM, and MJ contributed to the design and development of the study  
JH, IM, and NA contributed to data collection  
JH, IM, NA, SM, and MJ contributed data analysis and interpretation:  
JH, IM, and NA participated in the writing of the manuscript  
NA, SM, and MJ participated in the critical review  
JH, IM, NA, SM, and MJ provided approval for the final manuscript.

**Acknowledgements:** Sincere thanks to the patients who participated in the study.

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