



EARLY / OCCULT EHPVO NO LONGER IDIOPATHIC- UNRAVELLING METABOLIC CLUES IN A PROSPECTIVE STUDY IN ASIAN SCENERIO.

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Abstract

Background: Extrahepatic portal vein obstruction (EHPVO), characterized by occlusion and cavernomatous transformation of the portal vein resulting in portal hypertension. The clinical aspect of EHPVO is well established however its association with metabolic syndrome in adults has not been extensively studied. This study aims to explore the prevalence and components of metabolic syndrome in adult patients diagnosed with occult EHPVO.

Aims and Methods: In this prospective observational study conducted in a tertiary care center in Kashmir, India, 109 adult patients aged 15–75 years with non-cirrhotic, non-malignant EHPVO confirmed by abdominal ultrasonography were evaluated. Metabolic parameters such as body mass index (BMI), hypertension, diabetes or pre-diabetes, dyslipidemia, and hyperuricemia. Clinicoetiological evaluation along with follow-up outcomes, were recorded and analyzed. Statistical analysis was performed using SPSS. Categorical variables were expressed as frequencies and percentages and associations were assessed using the Chi-square test.

Results: Of the 109 patients, 56 (51.4%) were males and 53 (48.6%) females. Median age was 40.25 years. Metabolic evaluation showed normal BMI, dyslipidemia, pre-diabetes, hypertension, hyperuricemia in 78.9%, 27.5%, 14.7%, 30.3% and 11% respectively. Eighty percent (80%) were idiopathic, with substantial proportion of these patients displayed features of metabolic syndrome, suggesting a potential link between endothelial dysfunction and EHPVO.

Conclusion: This study highlights increased prevalence of metabolic syndrome in early occult EHPVO in Asian population thereby challenges the notion of idiopathic adult EHPVO. This condition however demonstrated a relatively benign course, with no mortality reported during the study's follow-up period.

Keywords: Extrahepatic portal vein obstruction (EHPVO), metabolic syndrome

Introduction.

Extrahepatic portal vein obstruction (EHPVO) is a primary vascular condition of the liver characterised by occlusion and cavernomatous transformation of the portal vein [1]. EHPVO has been found to be the commonest vascular cause of portal hypertension in non cirrhotic liver and which often goes unrecognised. Multiple tortuous hepatopetal collaterals, which develop around and inside the thrombus within the PV, known as cavernoma, appear within a span of 6–20 days following PV flow interruption [1]. In EHPVO, besides prothrombotic states, local factors leading to EHPVO in the absence of cirrhosis are pancreatitis, liver abscess, omphalitis, portal vein

phlebitis, umbilical vein catheterization (UVC), surgery around portal vein (splenectomy, cholecystectomy, Billroth-II procedure), and malignancies (pancreatic, hepatic, or duodenal) [2]. EHPVO has been seen to present very early as 6 weeks after birth or it may manifest in adulthood as well. Clinical presentation varies depends on onset of clinical disease and the age of presentation. Based on the location of the portal vein thrombosis, the EHPVO can be acute or chronic , complete or incomplete and also depends on the degree of extrahepatic portal venous system involvement. Variceal bleeding (70–80%), abdominal pain (36%), splenomegaly (20%), and altered metabolic features are the most prevalent clinical presentations in these patients [3]. This condition is predominantly recognized in children, yet its prevalence among adults [4] and its link with metabolic syndrome, has garnered recent attention. Although EHPVO in adults is frequently idiopathic, emerging studies suggest endothelial dysfunction could play a role, potentially tying EHPVO to metabolic syndrome components. This study explores the presence of metabolic syndrome and or its components and outcomes of adult patients presenting with EHPVO in the tertiary care centres of Kashmir, India.

Materials and Methods

A prospective observational study was conducted in a tertiary care hospital in Kashmir, Northern India. Patients aged 15 and 75 years diagnosed with extrahepatic portal vein obstruction (EHPVO) were enrolled. Patients with liver cirrhosis, prothrombotic conditions, Genetic mutations or malignancy were excluded from the study. Patients were referred to the outpatient department following a diagnostic abdominal USG, performed either for specific abdominal symptoms or as part of routine screening. For all patients with confirmed adult EHPVO, data were collected using a standardized proforma including demographic, clinical profiles, laboratory workup, findings from upper gastrointestinal endoscopy, liver function tests, viral markers, and potential risk factors for vascular occlusion. These risk factors included obesity, hypertension, diabetes, impaired fasting glycemia, dyslipidaemia, JAK2V617F mutation, other prothrombotic conditions, and antiphospholipid antibodies (APLA). Patients were followed-up from the time of enrolment. Patient underwent repeated endoscopies and ultrasounds over varying follow up period to monitor for complications of portal hypertension, such as oesophageal varices and splenomegaly. Statistical analysis was performed to examine associations between patients' etiological factors, clinical parameters, and outcomes. Categorical variables were expressed as frequencies and percentages, and associations were assessed using the Chi-square test. Ethical approval was obtained from the institutional review board.

Results

In this study of 109 adult patients with extrahepatic portal vein obstruction (EHPVO), the median age of presentation was +40.25 years with majority belonging to 25-35 years. The prevalence of metabolic syndrome components was assessed, focusing on obesity, dyslipidemia, hyperuricemia, hypertension, and diabetes or pre-diabetes. Most patients (86; 78.9%) had a normal BMI. Obesity was identified in 6 patients (5.5%), overweight in 14 patients (12.8%), and underweight in 3 patients (2.8%). Dyslipidaemia was present in 30 patients (27.5%). Hyperuricemia was observed in 12 patients (11%). Diabetes/Pre-diabetes was seen in 16 patients (14.7%) and Hypertension was seen in 33 patients (30.3%), as is shown in Figure 1. Most adult EHPVO belonged to the idiopathic group (87/109,80%). The etiological profile of EHPVO and its relationship with components of metabolic syndrome is shown in Figure 2. Patients were followed for up to 5 years, with all (100%) completing a 6-month follow-up. Long-term follow-up was achieved by 94.5% at 1 year, 61.5% at 2 years, 25.7% at 3 years, and 4.6% at 5 years. Most patients (96; 88.1%) did not require specific treatment. Outcomes varied based on the condition of those who did. No mortality was observed during the 5-year follow-up period, suggesting a favourable outcome for most patients, particularly those with metabolic syndrome components.

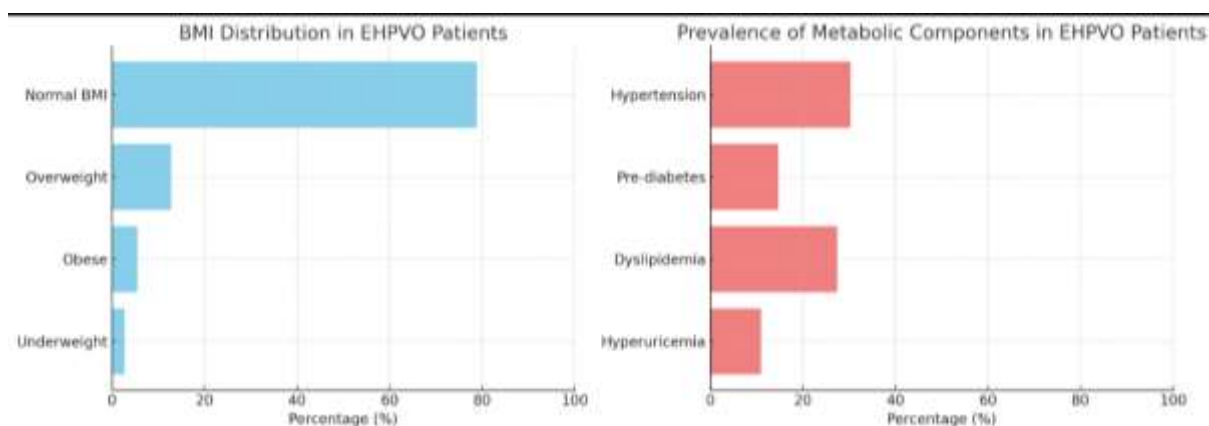


Figure 1.

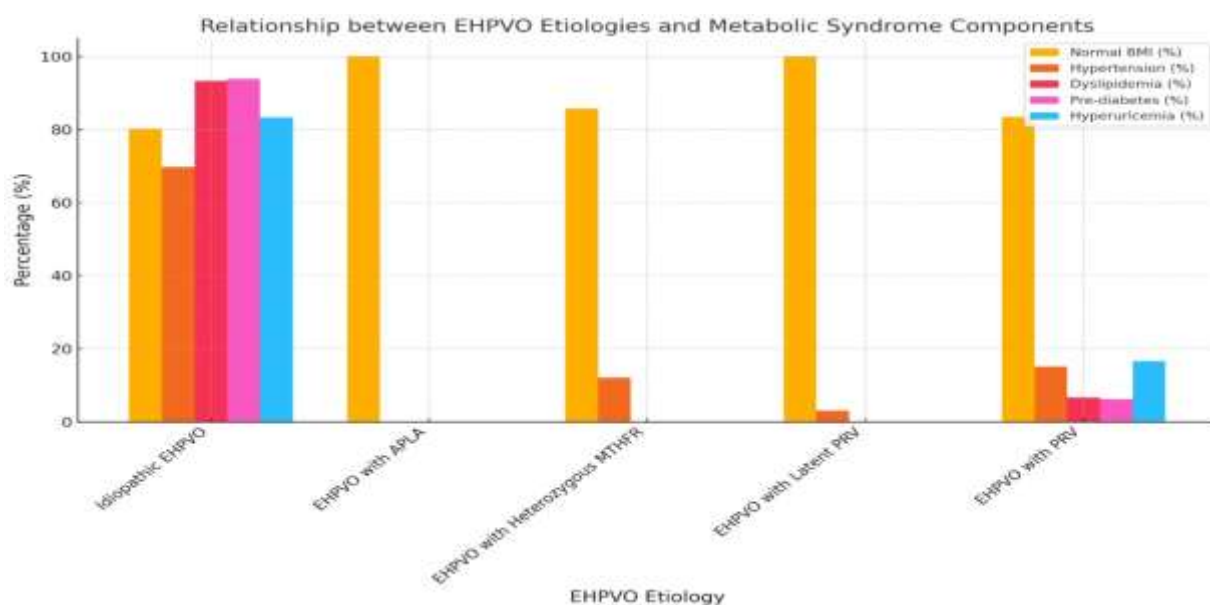


Figure 2.

Discussion

This study aimed to explore the presence and significance of metabolic syndrome and its components in adult patients with extrahepatic portal vein obstruction (EHPVO) in a tertiary care setting in Kashmir, India. We studied the demographic, clinical, and metabolic profile of adult EHPVO, otherwise associated with developing countries' pediatric populations [5,6]. The median age of EHPVO presentation in our study was 40.25 years. Similar findings have been seen in recent literature showing rising incidence among adults, especially in the absence of identifiable local precipitating factors [7]. In our cohort, 80% of cases were idiopathic, which underscores the need to investigate systemic and subclinical contributors, such as metabolic syndrome and endothelial dysfunction. A notable finding of our study was the high prevalence of individual components of metabolic syndrome in the EHPVO cohort—hypertension (30.3%), dyslipidemia (27.5%), and diabetes or pre-diabetes (14.7%). These proportions suggest a possible pathophysiological link between metabolic dysregulation and EHPVO. Although most patients had normal BMI, the presence of metabolic syndrome components in non-obese individuals may reflect a growing understanding that ectopic fat deposition and insulin resistance, rather than generalized obesity, drive vascular injury and thrombosis [8]. Endothelial dysfunction, a hallmark of metabolic syndrome, has been hypothesized as a mechanistic link in idiopathic EHPVO. Insulin resistance, oxidative stress, and chronic inflammation may predispose to a prothrombotic state via impairment in endothelial nitric oxide production and promotion of vascular stasis and thrombogenesis [9].

Additionally, dyslipidemia, especially elevated triglycerides and reduced HDL, may impair endothelial integrity and promote a hypercoagulable milieu [10].

The absence of mortality over a 5-year follow-up in our cohort is encouraging and consistent with prior observations that EHPVO, though a chronic condition, has a relatively favorable prognosis with appropriate surveillance and management of complications such as variceal bleeding [11]. Only 11.9% of patients required specific interventions, underscoring the role of conservative monitoring and individualized risk stratification in such cases.

The study's findings suggest that EHPVO in adults should prompt evaluation not only for traditional causes such as pancreatitis, infections, or surgery but also for metabolic abnormalities. The recognition of metabolic syndrome or its components may guide both etiological understanding and long-term management strategies, including cardiovascular risk reduction and thrombotic risk assessment. The increasing association between EHPVO and metabolic syndrome components observed in our study aligns with recent findings from other regions in Asia and Europe [12].

Our study has several strengths, including its prospective design and relatively long-term follow-up. However, it is limited by its single-center setting and lack of a control group. Further multicentric, controlled studies are warranted to explore the mechanisms linking metabolic dysfunction and EHPVO in adults.

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