

# Extrahepatic portal venous obstruction (EHPVO)

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## Abstract

Portal hypertension is defined as a free portal vein pressure in excess of the normal 5-10 mmHg. Portal hypertension represents a dynamic and intricate interaction between the compensatory mechanisms of the body and the prevalent pathology. In western countries, cirrhosis of the liver accounts for more than 90% cases of portal hypertension. In India, extrahepatic portal venous obstruction (EHPVO) is responsible for about one third cases of adults and more than half of the cases in children as a cause of portal hypertension.<sup>1,2</sup> This article represents a review of this topic.

## INTRODUCTION

Portal hypertension is defined as a free portal vein pressure in excess of the normal 5-10 mmHg. Portal hypertension represents a dynamic and intricate interaction between the compensatory mechanisms of the body and the prevalent pathology. In western countries, cirrhosis of the liver accounts for more than 90% cases of portal hypertension. In India, extrahepatic portal venous obstruction (EHPVO) is responsible for about one third cases of adults and more than half of the cases in children as a cause of portal hypertension.<sup>1,2</sup>

Noncirrhotic portal hypertension constitutes nearly 50% of all cases of portal hypertension in developing countries.<sup>2</sup> The two most common causes are non-cirrhotic portal fibrosis (NCPF) and extrahepatic portal vein obstruction (EHPVO). NCPF is characterized by the presence of a patent splenoportal axis demonstrable at ultrasonography or splenoportovenography and no evidence of cirrhosis on liver biopsy in a patient with portal hypertension. Such patients generally present with repeated well tolerated episodes of hematemesis and massive splenomegaly. EHPVO, on the other hand, is generally diagnosed in a young adult presenting with repeated hematemesis and evidence of occlusion in the main portal vein with portal cavernoma formation and normal histology of liver.<sup>2</sup>

Portal biliopathy refers to abnormalities of the extrahepatic, intrahepatic bile ducts and gall bladder wall in patients with portal hypertension. Portal biliopathy has been reported to occur in 80-100% of patients of EHPVO, 40% patients of NCPF and 30% patients of cirrhosis<sup>3,4</sup>. Paracholedochal and

paracholecystic collaterals either by compression or ischemic injury lead to indentation, caliber irregularity, angulation, strictures and ectasias of the bile ducts. Gall bladder varices are observed in 34% of patients of EHPVO, 24% of patients of NCPF and 13% patients of cirrhosis.<sup>5</sup> Presence of portal vein thrombosis is an important determinant for the development of gall bladder varices. Although, biliary abnormalities are common in portal hypertension, only a few patients present with jaundice, pain and cholangitis. Choleodocholithiasis has been reported to occur in 17% of patients with portal biliopathy.<sup>6</sup>

## REVIEW

Noncirrhotic portal hypertension is the portal hypertetion caused by pre-hepatic or intra-hepatic lesions in the absence of cirrhosis or venous outflow obstruction. It includes extrahepatic portal venous obstruction (EHPVO) and noncirrhotic portal fibrosis (NCPF). Other causes are, Schistosomiasis, Budd-Chiari syndrome, veno-occlusive disease and congenital hepatic fibrosis.<sup>3</sup>

In the western countries, cirrhosis accounts for more than 90% cases of portal hypertension and EHPVO constitutes less than 5% of cases. In India, noncirrhotic portal hypertension constitutes up to 50% of all patients of portal hypertension.<sup>7</sup>

EHPVO is an important cause of non-cirrhotic portal hypertension in third world countries. The most common site of block is at the portal vein formation [90%] and total block of splenoportal axis is seen in 10% of cases.<sup>8</sup> The etiology and clinical features are different in children and adults. In

children, the causes are umbilical sepsis, neonatal systemic sepsis, umbilical catheterization and developmental anomalies, other causes include dehydration, multiple exchange transfusions and sepsis. In adults, important causes are neoplastic diseases, infections, pancreatitis, myeloproliferative disorders and hypercoagulable states. The cause of portal vein block is obscure in 50% of cases. Loarroche et al, found that 40% of neonates having umbilical vein catheterization developed portal vein thrombosis after 24 to 48 hours and 100% after 3 days.<sup>9</sup> These thrombi resolve within short period of time. In another study by Thomson et al, no case of portal vein thrombosis was found in 470 neonates having umbilical vein catheterization.<sup>10</sup>

Myeloproliferative disorders are important cause of EHPVO in adults. Vella et al in their series reported latent myeloproliferative disorder in 58% of patients of EHPVO and 57% of these developed an overt disorder during follow up.<sup>11</sup> Underlying prothrombotic state is an important cause of portal vein thrombosis. Among them prothrombin gene mutation, factor V Leiden, protein C, protein S deficiency and antithrombin III deficiency are important. Cirrhosis itself is a predisposing factor for portal vein thrombosis. Portal vein thrombosis occurs in 0.6-7% of patients with cirrhosis.<sup>12</sup>

Other important causes of portal vein thrombosis in adults are neoplastic diseases. Pancreatic carcinoma and hepatocellular are important causes of portal vein thrombosis. Portal vein thrombosis may also be as a result of direct invasion of portal vein by tumor, extrinsic compression or periportal fibrosis following surgery or radio therapy. Infections that may lead to portal vein thrombosis include portal pyemia, biliary tract infections, post surgical sepsis, pancreatitis, diverticulitis and generalized septicemia.

Hemodynamic studies have shown that wedged hepatic venous pressure is within normal limits and intra splenic pressure is significantly elevated in patients with EHPVO.<sup>13</sup> Intra variceal pressure closely represents the portal pressure and intrasplenic pressure. The systemic vascular resistance is significantly lower and cardiac output is higher in patients with EHPVO. This is due to the extensive porto systemic venous collateral circulation.<sup>14</sup>

Braillon et al have shown that azygous blood flow is markedly increased in patients with presinusoidal portal hypertension and is significantly decreased by propranolol.<sup>15</sup>

The most common presentation of EHPVO in children is

well tolerated variceal bleed and splenomegaly. Ascites can occur in upto 13% of patients.<sup>16</sup> It is transient, usually following GI bleed or surgery. It is more frequent in adults than in children. Growth retardation can be present in children. In a prospective study by Sarin et al on 61 children of EHPVO, 50% of these patients had growth retardation and short stature. Hypersplenism manifested by thrombocytopenia and leukopenia was seen in 40-80% of patients. The hypersplenism does not correlate with splenic size. Ectopic varices are seen in a higher percentage of patients with EHPVO. Rectal varices has been reported to be present in 80% patients with EHPVO, 28% patients with cirrhosis and 30% patients with NCPF. Portal colopathy was seen in 40% patients of EHPVO, 14% of cirrhosis and 15% cases of NCPF. Overt bleeding was present in 8% cases of rectal varices and 4% cases of colopathy.<sup>17</sup>

Koshy et al in a retrospective analysis of 150 patients of EHPVO reported that one third had associated splenic vein thrombosis, two third bled before the age of five and one sixth bled before the age of three years. Splenomegaly was present in 10% of patients. Patients who presented with splenomegaly had first bleed after 4-5 years, later bleeding continued to occur at the rate of once per  $2 \pm 1.5$  years.<sup>21</sup>

### NON CIRRHOTIC PORTAL FIBROSIS (NCPF)

Non cirrhotic portal fibrosis (NCPF) has been reported from all over the world. NCPF has different names in different parts of the world. Its prevalence is 3-5% of all patients with portal hypertension.<sup>18</sup> In India the frequency is 15-18% of patients with portal hypertension.<sup>18,19</sup> Most series from different parts of India show male predominance with ratio of 2:1.<sup>20</sup> Koshy et al, described double peak incidence, one at 21-25 years and, another at 36-40 years. No definitive genetic predisposition has been found.<sup>21</sup> The relationship with arsenic has been suggested by many workers.<sup>22,23</sup> Increased hepatic arsenic has been attributed to high arsenic contents of drinking water, soil and vegetables. Sama et al suggested that repeated septic embolisation of the portal circulation and subsequent thrombophlebitis could be the cause of NCPF.<sup>24</sup>

Gastrointestinal hemorrhage is the most important symptom of NCPF and it occurs in 53-93% of the cases.<sup>25</sup> Bleeding episodes are generally well tolerated. The next symptom is the awareness of the lump in the left side of the abdomen.<sup>26,27</sup> Splenomegaly is universal. Ascites is present in 2-13% of patients.<sup>25</sup> The main splenoportal axis is patent and smaller branches of the portal vein are involved.

### PORTAL BILIOPATHY

Portal biliopathy refers to abnormalities of the extrahepatic bile ducts and intrahepatic bile ducts and gallbladder wall in patients with portal hypertension. These changes are most commonly seen in patients with extrahepatic portal vein obstruction. The changes include localized strictures or dilatations, irregular walls, angulation and displacement. The left hepatic duct is involved most commonly and severely, and this may be due to formation of prominent collateral veins where the umbilical vein joins the left branch of portal vein.<sup>4</sup> Sometimes these changes become severe enough to give rise to obstructive jaundice. Bile stasis may result in increased incidence of choledocholithiasis and cholelithiasis.

The development of portal hypertension leads to opening of numerous venous collaterals that decompress into systemic circulation. The common collaterals include esophageal and gastric varices. Varices due to paracholedochal and epicholedochal venous plexuses around CBD leads to protrusion of these veins in to lumen leading to compression of the lumen. Other possible mechanisms are vascular injury which leads to ischemia and fibrous stricture formation of the biliary system.

The earliest evidence of choledochal varices was first reported by Hunt in 1965.<sup>28</sup> Later on Meredith et al reported a few cases of common bile duct obstruction caused by extensive collateral venous circulation at the porta hepatis.<sup>29</sup> The cholangiographic changes caused by choledochal varices were first reported by William et al.<sup>30</sup>

In a study by Sarin et al, endoscopic retrograde cholangiopancreatographic (ERCP) changes were seen in 80% of the cases of EHPVO.<sup>3</sup> The ERCP changes in portalbiliopathy have been classified by Chandra et al.<sup>4</sup>

Type I - Involvement of only extrahepatic bile ducts

Type II - Involvement of only intrahepatic bile ducts

Type IIIa - Involvement of extrahepatic and unilateral (right or left) intrahepatic ducts

Type IIIb - Involvement of extrahepatic and bilateral intrahepatic ducts

### STUDIES ON ERCP

Dilawari et al studied twenty consecutive patients with the established diagnosis of extrahepatic portal venous obstruction. There were 16 men and four women. Successful ERCP was performed in all the 20 patients. The papilla and

pancreatic duct was normal in all these patients. Eighteen out of these 20 patients had abnormalities of midportion of the main bile duct. All 18 had indentations suggestive of external compression by choledochal varices. All patients had an abnormal looking left hepatic duct and its branches. These abnormalities were focal narrowing, dilatation, irregular walls and clustering of intrahepatic branches. Eleven of 20 patients had severe abnormalities and 9 had moderate changes. The abnormalities in right hepatic duct and its branches were similar but with a lesser degree of severity. The postulated mechanisms of biliary abnormalities in extrahepatic portal vein obstruction are either extrinsic compression by collaterals or ischemic injury due to venous thrombosis.<sup>31</sup>

Khuroo et al prospectively studied 21 consecutive patients with extrahepatic portal venous obstruction for evidence of biliary tract disease. Two patients were first diagnosed as extrahepatic cholestasis, another had recurrent cholangitis and another five had icterus on clinical examination. Liver function tests revealed elevated bilirubin levels in 14/21 (66.6%) patients, elevated alkaline phosphatase levels in 17/21 (80.9%), and elevated serum alanine transaminase levels in 8/21 (38%). Endoscopic retrograde cholangiography revealed abnormal findings in 17/21 (80.9%) patients. These changes involved the common bile duct (66.%) more often than they did the hepatic bile ducts (38.19%). Cholangiographic abnormalities included strictures (52.4%), caliber irregularity (23.8%) segmental upstream dilatation (42.8%), ectasia (9.5%) collateral veins causing extraluminal bile duct impressions (14.3%), and displacement of ducts (9.5%). The possible mechanism of these abnormalities were said to be collateral veins bridging the blocked portal vein, causing bile duct impressions, fibrous scarring of porta hepatis and ischemic injury to bile ducts, leading to stricture formation and caliber irregularity.<sup>32</sup>

Nagi et al evaluated cholangiographic abnormalities resulting from extrahepatic portal venous obstruction (EHPVO) by sonography and endoscopic retrograde cholangiopancreatography( ERCP). Forty three patients with an established diagnosis of EHPVO were subjected to duplex Doppler sonography and ERCP. Of these 8 patients had obstructive jaundice. Dilated common bile duct with pericholedochal varices was seen in five EHPVO patients with obstructive jaundice. ERCP revealed cholangiographic abnormalities in 40 patients (93%). Extrahepatic bile ducts were involved in 100% of cases compared to the intrahepatic

bile ducts (57%). Abnormalities noted were contour irregularity with indentations, displacement, angulation, strictures and filling defects in the extrahepatic ductal system. Intrahepatic bile duct showed dilatation with areas of narrowing and filling defects.<sup>33</sup>

Solomi et al have reported a case of cholestasis due to obstruction by portal cavernoma. Endoscopic retrograde cholangiography was done for palliative treatment of the stenosis of the common bile duct which was achieved by stenting.<sup>34</sup>

Malkan et al studied biliary changes due to portal hypertension. Cholangiopathy was detected by cholangiography in 17 of 20 patients with extrahepatic portal vein obstruction. Abnormalities were strictures and caliber irregularity in common bile duct (5), common hepatic duct (7), right hepatic duct (8) and left hepatic duct (11). One of the 11 patients with noncirrhotic portal fibrosis had a dilated hepatic ducts. Three of the 11 patients with cirrhosis had pruning in the intrahepatic ducts. Eight patients with portal vein obstruction had elevated serum alkaline phosphatase levels, two had elevated bilirubin levels. Ultrasonography detected gallbladder varices (11) and choledochal varices (9) in patients with extrahepatic portal venous obstruction.<sup>35</sup>

### STUDIES ON MRCP

Akaki et al reported two cases of bile duct stenosis due to portal cavernoma on magnetic resonance cholangiopancreatography. On contrast enhanced MR portography peribiliary tortuous vessels were evident, indicating portal cavernoma.<sup>36</sup>

### ASYMPTOMATIC BILIOPATHY

Bayraktar et al, studied biochemical consequences of cavernous transformation of the portal vein due to various etiologies. A total of 1247 patients with clinical evidence of portal hypertension were examined by using ultrasonography. 44 of these patients were found to have cavernous transformation of the portal vein (CTPV). CTPV was confirmed by splenoportography and digital subtraction angiography. Thirty five of these 44 patients had elevated serum bilirubin and alkaline phosphatase. Underlying disease in 7 of 44 was found to be Behcet's disease, congenital hepatic fibrosis in 5, congenital protein C deficiency in one and no etiology was found in remaining 26 patients. These patients were evaluated by ERCP (34 patients), percutaneous transhepatic cholangiography (1 patient) and CT in 19 patients. The surgical findings in 10

of these patients were reviewed in light of USG, portographic and ERCP findings. On ERCP and percutaneous transhepatic cholangiography, irregular indulating narrowing and nodular extrinsic defects were present in 33 of the 35 patients. It was concluded that mild elevation of direct reacting bilirubin and alkaline phosphatase occurred in CTPV associated with pseudocholangiocarcinoma sign. Presumably, these enzyme elevations are a result of compression of biliary tree by venous collaterals.<sup>37</sup>

### SYMPTOMATIC BILIOPATHY

Ouclin et al reported two cases of obstructive jaundice with markedly dilated collateral veins either in or around the bile duct in the setting of extrahepatic portal vein obstruction. In the first case proximal splenorenal shunt provided relief of biliary stenosis as well as eradication of esophageal varices. In the second case, obstructive jaundice was caused by probably cholangitis and was relieved by biliary drainage.<sup>38</sup>

Takehara et al, reported three cases which included one child and 2 adults, with unusual benign stricture of the extrahepatic bile duct. In 2 of these, stricture of the common bile duct was caused by compression of enlarged collateral varicose veins so called cavernous transformation following extrahepatic obstruction of portal vein was found and in another ischemic injury of CBD following right hepatectomy for mesenchymal hemartoma of the liver was the cause.<sup>39</sup>

Pickhardt et al, reported a case of dense portal vein calcification with secondary extrahepatic biliary stricture and cholangitis in an adult patient diagnosed as idiopathic portal hypertension at the age three. Biliary ductal dilatation proximal to echogenic shadowing near the portahepatis mimicking choledocholithiasis on sonographic examination. The obstruction was successfully managed with biliary stenting.<sup>40</sup>

Aguera et al, reported a case of cholestasis in a young patient with portal cavernomatosis. This clinical picture was as a consequences of extrinsic compression on the common bile duct by venous collateral.<sup>41</sup> Bejanin et al, reported 3 cases of portal cavernoma compressing the bile duct, one presented with asymptomatic cholestasis, and other two presented with obstructive jaundice and cholangitis.<sup>42</sup>

### OTHER PRESENTATIONS

Terada et al, reported recently the role of intrahepatic portal venous stenosis in the formation of hepatolithiasis. They

examined histologically the intrahepatic portal venous and arterial system in normal livers (n=13), extrahepatic biliary obstruction (n=18), intrahepatic biliary sludge and microcalculi (n=18) and stones (n=30). A scoring method was employed to quantify portal stenosis, portal phlebosclerosis, arterial stenosis and parenchymal atrophy. They found that these vascular changes were significantly more severe in patients with hepatolithiasis than in patients with biliary sludge and microcalculi or in extrahepatic biliary obstruction. There was no significant difference in the vascular changes except for arterial stenosis between the later two. There was positive correlation between vascular stenosis and parenchymal atrophy.<sup>43</sup>

Tanaka et al reported five cases of the portal vein obstruction (PVO) with intrahepatic stones. The coincidence rate of portal vein obstruction in intrahepatic stones was 5.8% (5 of 86 cases) and that of intrahepatic stones in portal vein obstruction was 45.5% (5 of 11 cases). All cases had one or more symptoms of cholangitis such as high grade fever, abdominal pain and jaundice prior to diagnosis of portal vein obstruction. The portal vein was occluded at the main trunk in 4 and in the left branch in one. Intrahepatic stones were found in bilateral hepatic lobes in 3 and in left lobe in 2. Numerous calcium bilirubinate stones were packed in the dilated intra hepatic bile ducts. Based on the results of the study, persistent cholangitis and gall stones were concluded cause for the development of portal vein obstruction accompanied by intrahepatic stones.<sup>44</sup>

### SHUNT SURGERY IN PORTAL BILIOPATHY

The postulated mechanisms of biliary abnormalities in EHPVO are extrinsic compression by collaterals and ischemic injury due to venous thrombosis. In a study by Dhiman et al, five patients of EHPVO with proved biliopathy on ERCP were subjected to shunt surgery. The post shunt surgery ERCP showed partial reversal of biliary abnormalities in three patients, complete reversal in one and no reversal in one. It was concluded that shunt surgery resulted in regression of some of the biliary abnormalities and relieved biliary obstruction, suggesting mechanical compression by collaterals as mechanism behind biliary abnormalities in EHPVO.<sup>45</sup>

Chaudhary et al studied nine patients of EHPVO with symptomatic portal biliopathy. Eight patients presented with jaundice, two had abdominal pain and one cholangitis. ERCP revealed abnormalities of the bile duct wall with strictures in 8 patients and bile duct calculi in two. Portal

systemic shunting relieved jaundice in 5 of the seven patients.<sup>46</sup>

**Gall bladder varices :** Gall bladder varices are portosystemic shunts between the cystic vein branch of portal vein either to systemic anterior abdominal wall veins or to patent portal vein branches within the liver. The prevalence of gall bladder varices as reported by West et al is 12% in patients with portal hypertension<sup>47</sup>. The prevalence of gall bladder varices as reported by Chawla et al is 30% in association with portal vein thrombosis.<sup>5</sup> The gall bladder varices showed no correlation with the site of block in splenoportal axis or variceal obliteration.

### GALL BLADDER FUNCTION IN PORTAL HYPERTENSION

Chawla et al studied 102 patients with portal hypertension (cirrhosis 38, NCPF 29 and EHPVO 35) and 25 healthy controls. Prevalence of gallbladder varices in cirrhosis, NCPF and EHPVO was 7%, 22%, and 34% respectively. The gallbladder residual volume (RV) and fasting volume (FV) were not different in these groups of patients compared with controls. Ejection fraction was similar in patients with or without gall bladder varices. It was concluded that gall bladder varices caused some gallbladder stasis but did not impair gallbladder function.<sup>5</sup>

### ENDOSCOPIC ULTRASOUND IN GALL BLADDER VARICES

Palozzi et al studied biliary varices on endoscopic ultrasonography (EUS) in 21 patients of EHPVO. These varices which were not visible on conventional imaging methods (CT and USG) were identified by EUS. In 16 patients (76%), they were in the wall of CBD, in 11 patients (52%) surrounding the CBD and in 9 (43%) they were in gall bladder. These varices were the cause of obstructive jaundice in three of the 21 patients. Two of these patients were treated using portosystemic shunting and other received biliary endoprosthesis.<sup>48</sup>

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