Non-cirrhotic Portal Hypertension

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KEYWORDS

- Portal hypertension • Non-cirrhotic portal fibrosis
- Extrahepatic portal venous obstruction • Endotherapy • Shunt surgery
- Portal biliopathy

KEY POINTS

- NCPH includes a wide range of disorders presenting with PHT, preserved liver synthetic functions and normal or mildly elevated hepatic venous pressure gradient (HVPG).
- NCPF/IPH and EHPVO are two distinct diseases – former is a disorder of young adults, whereas later is a disease of childhood.
- Likely pathogenesis in both of them relates to recurrent infections in a prothrombotic individual.
- Diagnosis needs exclusion of cirrhosis in NCPF/IPH and presence of a cavernoma in EHPVO.
- Effective management focused on PHT results in good long term survival.

Non-cirrhotic portal hypertension (NCPH) encompasses a wide range of vascular conditions leading to portal hypertension (PHT) associated with normal or mildly elevated hepatic venous pressure gradient (HVPG), whereas the portal venous pressure gradient between the portal vein (PV) and inferior vena cava is comparable or higher than cirrhotic PHT. The diseases leading to NCPH are classified anatomically by the site of resistance to blood flow as prehepatic, hepatic, and posthepatic; hepatic causes are further subdivided into presinusoidal, sinusoidal, and postsinusoidal.[1–3] **Fig. 1** gives an approach and classifies various disorders listed under the category of NCPH. In most of the conditions leading to NCPH, PHT is a late manifestation of the primary disease, except for NCPF and extrahepatic PV obstruction (EHPVO) whereby PHT is the only or predominant manifestation.[1,3,4] The present review describes these 2 entities in detail with a brief discussion on some other causes. Some of the contrasting differences between NCPF and EHPVO have been highlighted in **Table 1**.

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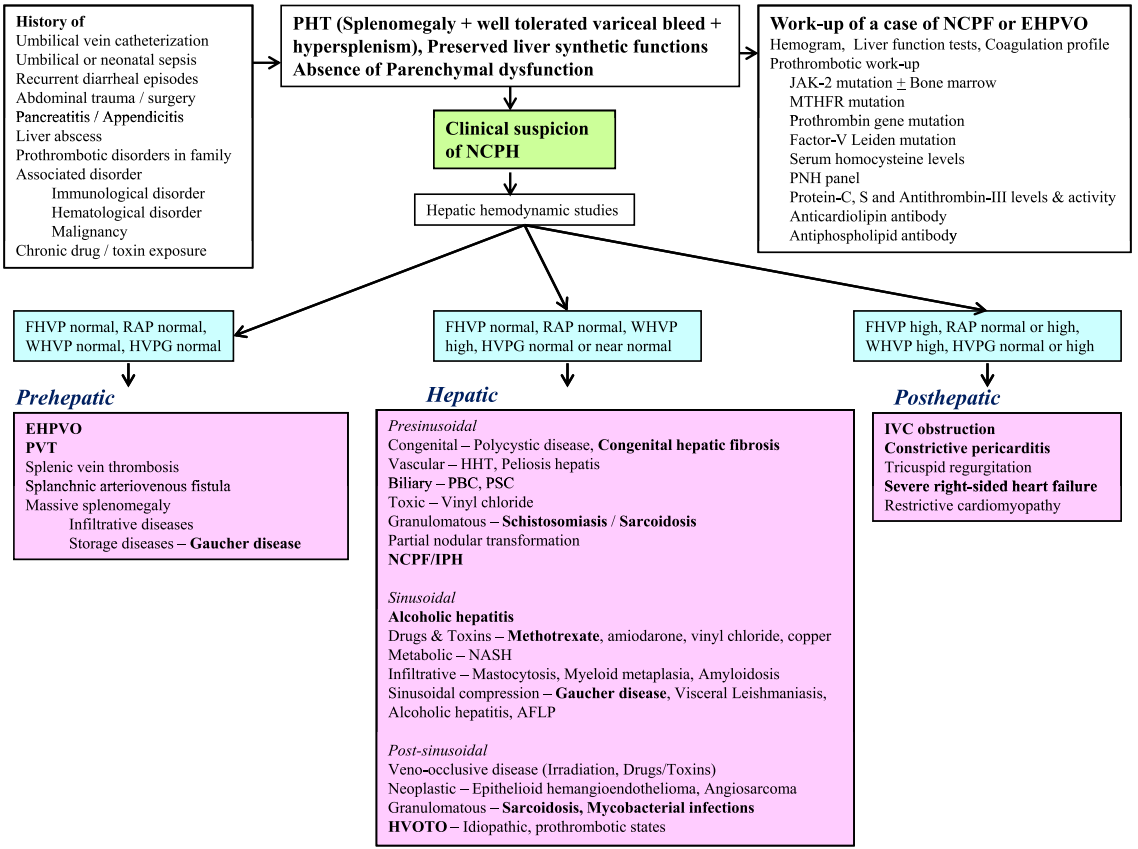

Fig. 1. Approach to a patient of NCPH. NCPH is suspected on clinical grounds and various causes are then classified by hepatic hemodynamic studies as shown. AFLP, acute fatty liver of pregnancy; EHPVO, extrahepatic portal venous obstruction; FHVP, free hepatic venous pressure; HHT, hereditary hemorrhagic telangiectasia; HVOTO, hepatic venous outflow tract obstruction; IPH, idiopathic portal hypertension; IVC, inferior vena cava; JAK-2, Janus kinase-2; MTHFR, methylene tetrahydrofolate reductase deficiency; PBC, primary biliary cirrhosis; PNH, paroxysmal nocturnal hemoglobinuria; PSC, primary sclerosing cholangitis; PVT, portal vein thrombosis; RAP, right atrial pressure; WHVP, wedge hepatic venous pressure. (*Data from* Refs.[1,3,26])