

A multisociety consensus statement on a new common definition and diagnostic criteria for PSVD or NCPF.

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Noncirrhotic portal hypertension has historically been described using heterogeneous and region-specific terminology-such as idiopathic portal hypertension (IPH), noncirrhotic portal fibrosis (NCPF), obliterative portal venopathy, and nodular regenerative hyperplasia-leading to substantial variability in diagnosis, reporting, and international research collaboration. Differences in guideline definitions from major societies (AASLD, EASL, and APASL), together with the presence of characteristic histologic lesions in patients without clinically overt portal hypertension, have further complicated disease classification. To address these challenges, a large, multisociety, international initiative was convened to harmonize nomenclature and diagnostic criteria. Representatives from liver, pathology, and pediatric hepatology societies across the Americas, Europe, and Asia participated in a structured consensus process that included specialized working groups and external Delphi validation. The initiative produced a globally harmonized and implementable diagnostic framework. Consensus was reached that the terms porto-sinusoidal vascular disorder (PSVD) and NCPF may be used interchangeably when identical diagnostic criteria are applied, and that they should be written as PSVD or NCPF. The diagnosis was defined as fundamentally clinicopathological, requiring integrated assessment. Core principles include the need for a high-quality liver biopsy (≥ 10 mm), mandatory exclusion of cirrhosis, and systematic exclusion of specific alternative conditions. Importantly, the consensus recognizes that PSVD or NCPF may be diagnosed even without clinical portal hypertension and may coexist with other liver diseases, provided cirrhosis is excluded. Standard-ized major and minor histologic criteria were developed collaboratively by expert pathologists and externally validated. Features of portal hypertension were harmonized into specific and nonspecific categories applicable to routine clinical practice. An integrated diagnostic scoring system incorporating histology, clinical features, associated conditions, and concomitant etiologies was developed and validated using the Delphi method. This consensus provides the first internationally endorsed, unified framework for the diagnosis of PSVD or NCPF. Its global implementation is expected to reduce diagnostic variability, improve comparability across regions, and facilitate the development of robust, internationally harmonized clinical and translational research cohorts.