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Porto-Sinusoidal Vascular Disease: A Concise Updated Summary of Epidemiology, Pathophysiology, Imaging, Clinical Features, and Treatments

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Take-home points

- Porto-sinusoidal vascular disease (PSVD) is a rare disorder usually characterized by signs of portal hypertension in the absence of an identifiable etiology, such as cirrhosis, and is largely under-recognized because of insufficient disease awareness among physicians.
- The prevalence of PSVD varies geographically, and it is more commonly reported in developing countries than in developed countries.
- Imaging findings suggestive of PSVD include the absence of liver surface nodularity, hypertrophy of segments IV and I, and normal or mildly elevated liver stiffness values in patients with signs of portal hypertension.
- Liver biopsy is mandatory for the diagnosis of PSVD, and specific histologic signs include obliterative portal venopathy, nodular regenerative hyperplasia, and incomplete septal fibrosis.
- The treatment of PSVD focuses on managing portal hypertension-related complications and adopts the same strategy as that for patients with cirrhosis.
- The long-term prognosis of PSVD, which is generally better than that of cirrhosis, is associated with age, specific signs of portal hypertension, including ascites, and underlying conditions.

INTRODUCTION

Porto-sinusoidal vascular disease (PSVD) is a recently proposed nomenclature, which is loosely regarded as a disease entity of portal hypertension with no histological evidence of cirrhosis. Historically, Banti first described patients with anemia and splenomegaly without hematologic disorders, henceforth called "Banti's syndrome," a condition previously considered a primary splenic disorder with subsequent changes in the liver, with endophlebitis as a common etiology [1,2]. The paradigm shift happened in 1934, when McMichael first linked pathological alterations in the portal veins to portal hypertension in patients with "hepatolienal fibrosis," i.e., splenomegaly without cirrhosis [3]. Subsequently, various terms, including "idiopathic portal hypertension," "noncirrhotic portal fibrosis," "hepatoportal sclerosis," "incomplete septic cirrhosis," or "regenerative nodular hyperplasia," were used to name the abnormal manifestations until 2011 when the unifying term "idiopathic non-cirrhotic portal hypertension (INCPH)" was proposed by Schouten et al. [4]. Despite the more refined diagnostic criteria for ICHPH recently proposed by the European Association for the Study of the Liver [5], the definition of "INCPH" had several limitations in describing PSVD as currently understood. It excluded PSVD patients with 1) no portal hypertension due to early stages of the

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disease, 2) portal vein thrombosis that frequently occurs during the disease course, and 3) other coexisting causes of liver diseases. To overcome these limitations and cover all aspects of this rare disorder, the new term "PSVD" has been suggested by the Vascular Liver Disease Interest Group [6]. Nevertheless, awareness of PSVD remains rather low even among experts, leading to misdiagnoses or delayed diagnoses. This article aims to provide an updated account of the clinical, radiological, and pathological aspects of PSVD and raise awareness of this under-recognized disease.

Epidemiology

The prevalence of PSVD varies geographically, is reportedly higher in developing countries [7], and appears to be decreasing overall. Nevertheless, it accounts for about 15%–34% of all cases of portal hypertension [8-11]. In Japan, the incidence of INCPH (i.e. PSVD with portal hypertension) has dramatically reduced from 31% new cases of portal hypertension in 1975 to 11 new cases/year [12]. In Western countries, PSVD with portal hypertension is less prevalent, accounting for 3%–6% of all cases of portal hypertension [13-16]. PSVD is more prevalent in men in India and the West, whereas it is more common in women in Japan [13,16-19]. The average age of onset is significantly younger in India (25–35 years) than in the West (nearly 40 years) and Japan (43–56 years) [17-19]. INCPH can also occur in children [20]. In Korea, data remain limited on PSVD epidemiology, except for a few case series and reports. A recent study reported that the mean age at diagnosis was 46.3 years, and the proportion of men diagnosed was slightly higher (55.8%) [21].

A survey on PSVD awareness conducted in June 2022 among 95 Korean doctors (including residents [68.4%], clinical fellows [13.7%], and experts [17.9%] in gastroenterology, radiology, and pathology) found that approximately 60%–80% of the respondents did not know about the clinical manifestation, pathological and imaging characteristics, or treatment options for PSVD (Fig. 1). Such an insufficient disease awareness may lead to misdiagnosis or delayed diagnosis in a large number of cases. In fact, an earlier study found that 72.1% of PSVD patients were initially misdiagnosed with cirrhosis, and among those patients, it took a median time of 32 months for PSVD to be diagnosed [21].

Pathophysiology

The pathophysiology of PSVD remains poorly understood. An early study on portal hemodynamics in INCPH identified

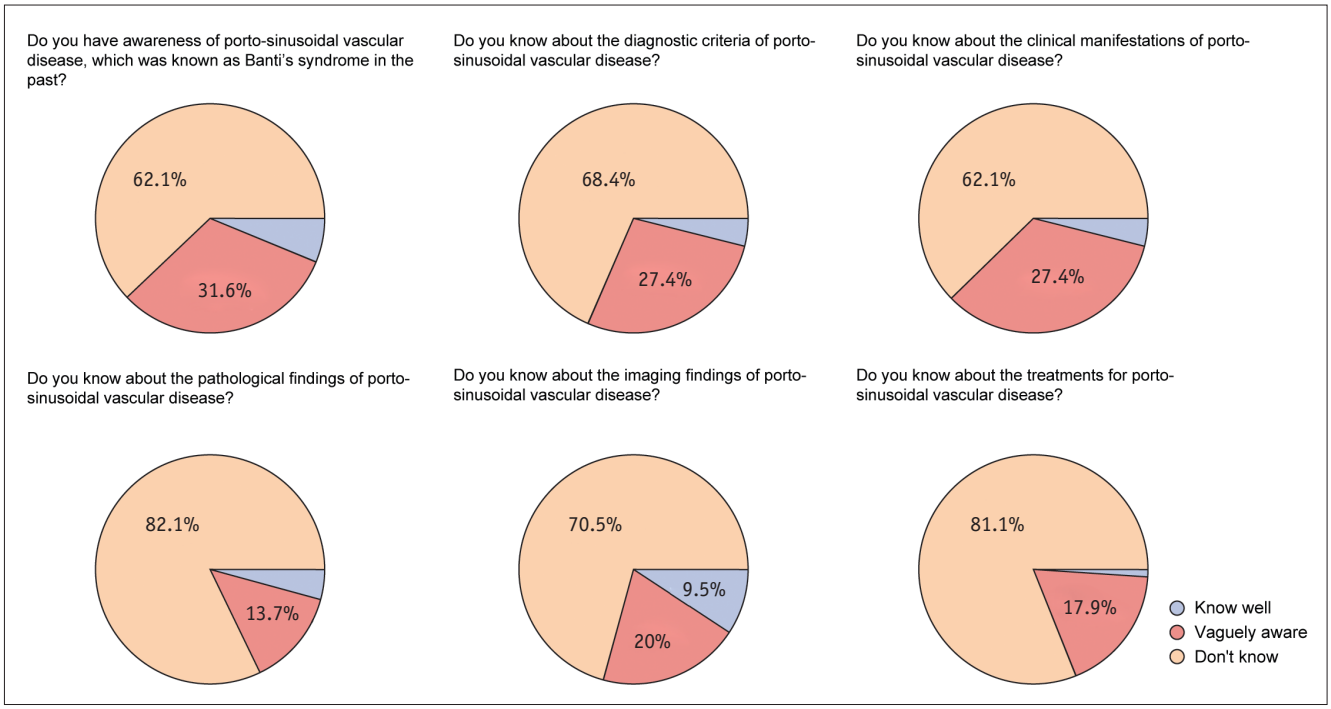

Fig. 1. Results from the awareness survey for porto-sinusoidal vascular disease. The survey was conducted in June 2022 among 95 Korean doctors in gastroenterology, radiology, and pathology.

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Korean Journal of Radiology
KJR

two distinct groups of patients with INCPH: one with markedly increased splenic and portal venous flow and moderately elevated portal vein pressure and the other with substantially increased portal vascular resistance and portal vein pressure [22]. On the basis of these findings, a dual hypothesis implicating both increased splenic blood flow and obliteration of small and medium portal venous branches was proposed (Fig. 2). In the first scenario, diffuse, high expression of inducible nitric oxide (NO) synthetase and endothelial NO synthetase have been observed in the sinus-lining cells of the spleen. The resulting intense NO release causes splenic sinus enlargement and splenomegaly, which in turn leads to increased splenic venous flow and an increase in portal pressure. In the second scenario, obliterative portal venopathy is the histological hallmark of PSVD. This obliteration of the portal venous microcirculation results in elevated intrahepatic resistance [4]. According to this hypothesis, diseases that can cause injury to the small or medium portal venous branch are likely to be potential causes of PSVD. In fact, associated conditions, including immune disorders, hematological diseases, prothrombotic conditions, HIV infection, recurrent gastrointestinal infections, congenital familial defects (i.e., mutations in the telomerase gene complex, *KCNN3*, or *DGUOK*, Adams–Oliver syndrome, Turner's syndrome, familial obliterative portal venopathy, and cystic fibrosis), and drug exposure, have been identified in 43%–58% of patients with PSVD [6,17,18,23]. This highlights the importance of ruling out systemic disorders associated with PSVD, using tests such as complete blood count, liver tests, serology for HIV infection, and a complete thrombophilia study, among others.

Diagnosis

PSVD is diagnosed on the basis of the absence of cirrhosis and presence of histological changes, with or without portal hypertension. Thus, liver biopsy is mandatory for the diagnosis of PSVD. For PSVD to be diagnosed, there must be at least one specific clinical or pathological sign, or one non-specific clinical sign and one non-specific pathological sign in combination (Table 1) without histological evidence of cirrhosis [6]. By definition, PSVD can be diagnosed without clinical signs of portal hypertension when PSVD-specific histological alterations such as obliterative portal venopathy, nodular regenerative hyperplasia, incomplete septal fibrosis, or cirrhosis are observed. Obliterative portal venopathy is characterized by an increase in the connective tissue around the portal veins, irregular thickening of the vessel walls, and an eccentric narrowing of the vessel lumen, which can lead to complete occlusion of the veins. Nodular regenerative hyperplasia is characterized by the transformation of normal hepatic parenchyma into tiny nodules, which are typically 1–3 mm in size and often macroscopically paler than normal parenchyma. Finally, incomplete septal fibrosis or cirrhosis is identified by the presence of incomplete, thin, perforated, or blind-ended septa that intermittently delimit rudimentary nodules.

Unlike INCPH, the presence of chronic liver disease,

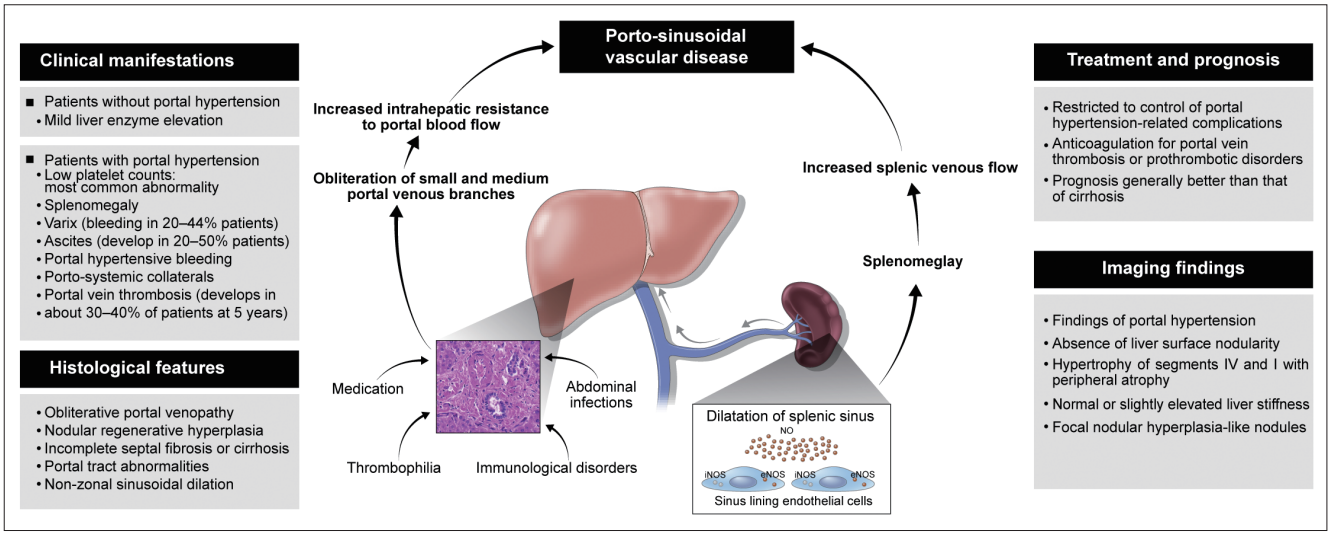

Fig. 2. Hypothesized pathophysiology and disease characteristics of porto-sinusoidal vascular disease. eNOS = endothelial nitric oxide synthase, iNOS = inducible nitric oxide synthase, NO = nitric oxide

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such as alcoholic liver disease, non-alcoholic fatty liver disease, or viral hepatitis, does not rule out the diagnosis of PSVD when PSVD is suggested by histology. The presence of extrahepatic portal vein thrombosis, which frequently occurs during the course of this disease entity, does not exclude PSVD either. However, the presence of liver-related hepatic vein obstruction and specific causes of microvascular disease, such as Budd–Chiari syndrome, sinusoidal obstruction syndrome, hepatic schistosomiasis, cardiac failure or Fontan surgery, Abernethy syndrome, hereditary hemorrhagic telangiectasia, sarcoidosis, or congenital hepatic fibrosis, exclude PSVD [6].

Imaging

Although the imaging features of PSVD are not fully specific for accurate diagnosis, they may help differentiate it from cirrhosis. Imaging findings of PSVD include common features of portal hypertension such as splenomegaly and portosystemic collaterals. The pre-symptomatic phase of PSVD without portal hypertension would be unrecognizable on imaging. The absence of surface nodularity in patients with signs of portal hypertension can be an initial clue to suspect PSVD rather than cirrhosis (Fig. 3A) [24]. While the presence of segment IV atrophy in combination with segment I hypertrophy suggests cirrhosis, a combination

Table 1. Clinical and Pathological Signs for the Diagnosis of Porto-Sinusoidal Vascular Disease

	Clinical Signs	Pathological Signs
Specific	Varix	Obliterative portal venopathy
	Portal hypertensive bleeding	Nodular regenerative hyperplasia
	Porto-systemic collaterals	Incomplete septal fibrosis or cirrhosis
Not specific	Ascites	Portal tract abnormalities (multiplication and dilatation of arteries, periportal vascular channels, and aberrant vessels)
	Thrombocytopenia	Architectural disturbance (irregular distribution of the portal tracts and central veins)
	Splenomegaly	Non-zonal sinusoidal dilatation
		Mild perisinusoidal fibrosis

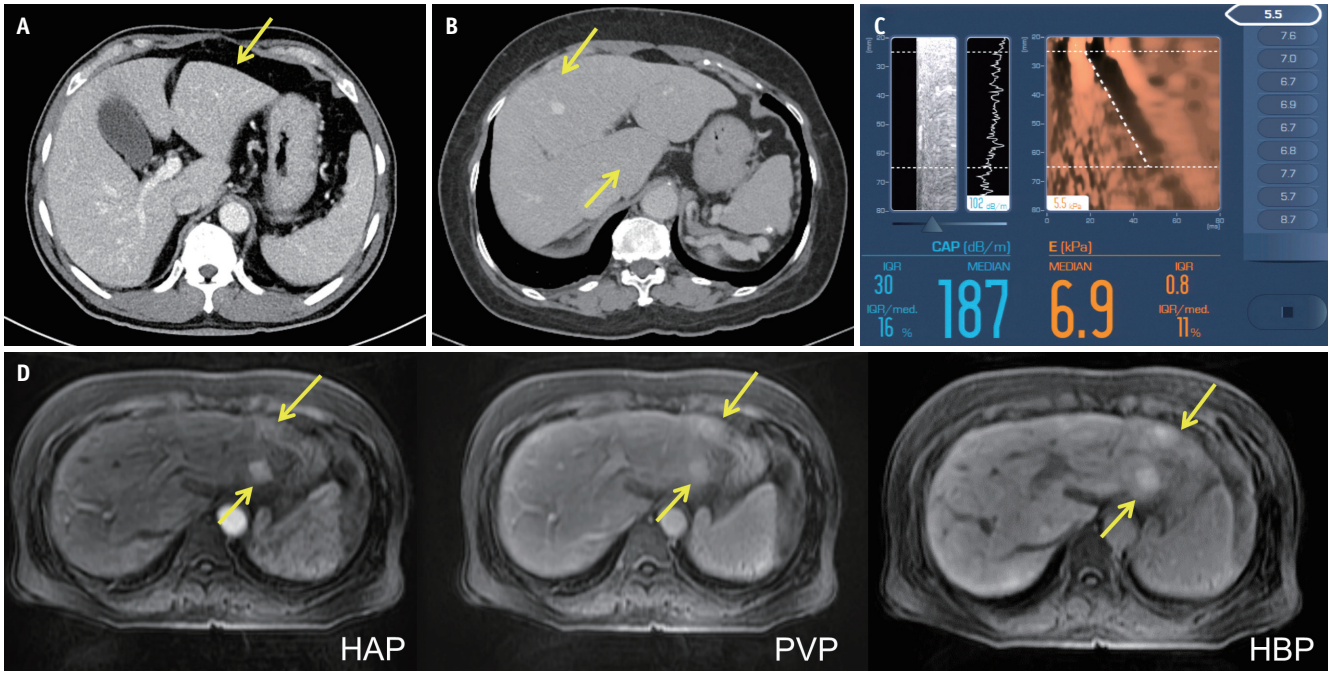

Fig. 3. Representative imaging features of porto-sinusoidal vascular disease.
A. Absence of liver surface nodularity (arrow) is noted on CT. **B.** Hypertrophy of segments IV and I (arrows) with peripheral atrophy are noted on CT. **C.** Slightly elevated liver stiffness value of 6.9 kPa according to a transient elastography. **D.** Focal nodular hyperplasia-like nodules (arrows) showing arterial phase hyperenhancement, lack of portal venous washout, and hyperintensity on the HBP of hepatobiliary agent-enhanced MRI. HAP = hepatic arterial phase, HBP = hepatobiliary phase, PVP = portal venous phase

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Korean Journal of Radiology
KJR

of segment IV and I hypertrophy with peripheral atrophy may indicate abnormal portal venous inflow such as PSVD or portal vein thrombosis (Fig. 3B) [25,26]. Intrahepatic and/or extrahepatic portal vein abnormalities, including portal vein thrombosis, are more frequently observed in PSVD than in cirrhosis [21,24,25]. The heterogeneity of parenchymal enhancement on the arterial and venous phases of dynamic CT is more frequently observed in PSVD than in cirrhosis [21,25]. Parenchymal enhancement in the hepatobiliary phase of hepatobiliary agent-enhanced MRI (HBA-MRI) is greater in patients with PSVD than in those with cirrhosis. Moreover, the portal hypertension grade tends to be higher [21].

In PSVD, liver stiffness values are usually normal or only slightly elevated (Fig. 3C) [27,28]. Thus, the clinical signs of portal hypertension of unknown etiology and normal or only mildly elevated liver stiffness values strongly support the diagnosis of PSVD. In contrast, spleen stiffness is increased in PSVD. Thus, the spleen-to-liver stiffness ratio is significantly higher in patients with PSVD than in patients with cirrhosis [29].

Hepatic vein venography has revealed hepatic venovenous communications in > 50% of patients with PSVD, which explains the difficulty of measuring a proper wedge pressure or the typically normal or only slightly elevated hepatic venous pressure gradient (< 10 mm Hg) in most PSVD patients [27,30,31]. A venography finding supports the clinical suspicion of PSVD.

Focal nodular hyperplasia-like nodules are the most commonly observed focal hepatic lesions in patients with PSVD and may be confused with hepatocellular carcinoma. Typical imaging findings of focal nodular hyperplasia-like nodules in patients with PSVD included arterial phase hyperenhancement with no portal venous washout and hyperintensity on the hepatobiliary phase of HBA-MRI (Fig. 3D) [21]. Half of the cases were observed as multiple lesions, and in these cases, the size varied from < 1 cm to 2.7 cm [21]. Estimates for the development of hepatocellular carcinoma in patients with PSVD differed widely. No malignant lesions developed in a previous cohort of 43 patients with PSVD during a median follow-up of 46 months [21], whereas two patients developed hepatocellular carcinoma during a median follow-up of 37 months in another cohort of 91 patients with PSVD [32]. However, more than 30% of the latter cohort included patients with risk factors for hepatocellular carcinoma, such as hepatitis B or C virus infection.

Clinical Presentation

The most notable difference between the newly suggested PSVD and previous INCPH is that PSVD diagnosis does not necessitate any signs of portal hypertension. Obliterative portal venopathy in liver biopsy could be observed in an early, pre-symptomatic phase of PSVD without portal hypertension [33]. In this clinical setting, PSVD without portal hypertension is often diagnosed by liver biopsy in patients with mild liver enzyme elevation after the exclusion of a specific cause. Although data on PSVD without portal hypertension are insufficient, the condition is thought to be more common than previously assumed (19% of patients with cryptogenic liver disease) [6].

In patients with PSVD and signs of portal hypertension, the most common laboratory finding is a low platelet count. Normal or slightly elevated levels of liver enzymes such as alanine aminotransferase or alkaline phosphatase, usually less than twice the upper limit of normal values, could be observed in over 80% of patients with PSVD with signs of portal hypertension. Two-thirds of patients with PSVD with portal hypertension had large varices, and 20%–44% of patients presented with variceal bleeding at initial presentation. Twenty percent of patients developed large varices within an average of 10 years after diagnosis [18]. Ascites developed in 20%–50% of the cases with PSVD with portal hypertension, with a triggering event occurring in more than half the cases [17,18]. At 5 years after the diagnosis of PSVD with portal hypertension, approximately 30%–40% of patients developed portal vein thrombosis [17-19]. Some data indicate that portopulmonary hypertension, hepatopulmonary syndrome, and liver regenerating nodules can occur in patients with PSVD, similar to other vascular diseases [17]. However, the risk factors for these complications are largely unknown.

Treatment and Prognosis

Currently, no PSVD-specific treatments to prevent disease progression exist. Instead, the treatment of associated systemic conditions or cessation of drugs that cause PSVD should be considered. In patients with PSVD and signs of portal hypertension, the current recommendation is to treat complications associated with portal hypertension, which is adopted from the guidelines for patients with cirrhosis [34]. The mainstay of the treatment for varices is endoscopic band ligation and/or beta-blockers, including propranolol

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and carvedilol. For patients with PSVD with ascites, a low-salt diet and diuretics should be considered. For patients with severe complications of portal hypertension, transjugular intrahepatic portosystemic shunts can be considered, with outcomes comparable to those in patients with cirrhosis [35]. Patients with severe hypersplenism may consider splenectomy or partial splenic embolization. These interventions have been shown to increase platelet counts [35,36]; however, their advantages have never been shown to exceed their hazards. Thus, these interventions should be reserved for rare patients with intractable symptoms associated with hypersplenism, such as repeated splenic infarcts or spontaneous bleeding events. Liver transplantation can be considered based on the same indications as those for cirrhosis. Data on the outcome or risk of recurrence after liver transplantation in patients with PSVD are sparse. At our institution, 30 patients who underwent liver transplantation between 2008 and 2021 showed a favorable outcome, with a 5-year survival rate of 96.7% without definite recurrence.

Another controversial issue in the management of PSVD is the role of anticoagulants. Several lines of evidence support the use of anticoagulants for PSVD. Portal vein thrombosis is a common complication, observed in 13%–45% of patients with PSVD during follow-up [17-19,37]. It may be recommended that the patency of the entire splanchnic circulation is evaluated at the time of diagnosis and then twice-yearly during follow-up [34]. PSVD is frequently associated with underlying prothrombotic conditions. However, because there are insufficient data showing the benefits of prophylactic anticoagulation, current expert opinions suggest considering anticoagulation only when a prothrombotic disorder is diagnosed or portal vein thrombosis has developed [12].

The overall long-term outcome of PSVD is generally better than that of cirrhosis, presumably because liver function can be preserved in the majority of patients [3]. Blood levels of bilirubin, prothrombin, and albumin were observed to change slightly on average over a follow-up period of 7–8 years in a cohort of 69 patients with PSVD and portal hypertension. This suggests that the disease is non-progressive or slowly progressive in most patients [17]. Based on an average follow-up of approximately 8 years, mortality of patients with PSVD has been observed to be between 15%–20% [17,18,23,32,38]. Prognostic factors for PSVD are not well established. Age, specific signs of portal hypertension including varices and ascites, and underlying conditions associated with PSVD have been associated with the risk of death [19,32].

CONCLUSIONS

PSVD is probably under-recognized and underestimated in Korea and perhaps elsewhere too due to insufficient awareness of the disease among physicians and insufficient familiarity with the imaging and histological features of PSVD among radiologists and pathologists, respectively. We recommend that physicians consider this condition when they encounter a clinical setting characterized by signs of portal hypertension with normal or slightly elevated liver stiffness and portal pressure values. Radiologists should be familiar with the imaging features of PSVD that provide clues for its suspicion. At present, liver biopsy is mandatory for the diagnosis of PSVD if clinical suspicion is raised. Thus, pathologists should be aware of the histological hallmarks of PSVD, particularly in situations in which the role of histology in the diagnosis is more complicated than simply ruling out cirrhosis. In several areas, including the pathophysiology of PSVD and treatments that can modify the natural course of the disease, our current understanding of PSVD has large gaps, and addressing them will require collaborative research.

Key words
Idiopathic noncirrhotic portal hypertension; Hepatoportal sclerosis; Obliterative portal venopathy; Disease awareness

Availability of Data and Material
The datasets generated or analyzed during the study are available from the corresponding author on reasonable request.

Conflicts of Interest
The authors have no potential conflicts of interest to disclose.

Author Contributions
Conceptualization: all authors. Funding acquisition: Won-Mook Choi. Investigation: all authors. Methodology: all authors. Project administration: Won-Mook Choi. Resources: Won-Mook Choi. Software: Won-Mook Choi. Supervision: Won-Mook Choi. Visualization: Won-Mook Choi. Writing—original draft: all authors. Writing—review & editing: Won-Mook Choi.

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