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Porto-sinusoidal vascular disease

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What is porto-sinusoïdal vascular disease?

The term porto-sinusoidal vascular disorder (PSVD) encompasses a wide spectrum of liver disorders, including the so-called non-cirrhotic portal hypertension, hepato-portal sclerosis, obliterative portal venopathy, nodular regenerative hyperplasia or hepato-portal sclerosis. These disorders are not strictly identical, but they have two main points in common: a) they affect the small (microscopic) vessels of the liver (Figure 1) and b) they can lead to portal hypertension. The prevalence of PSVD is unknown, but it is considered as a rare disease..

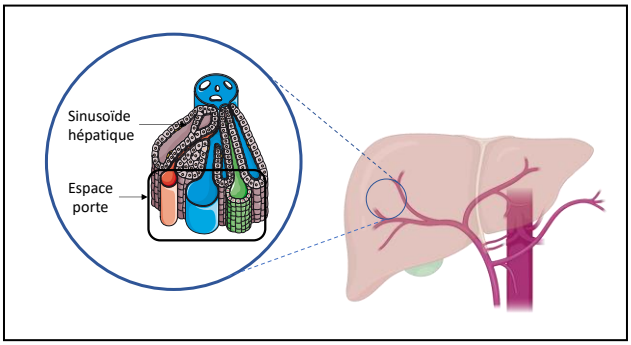

Figure 1: Schematic representation of the anatomy of the liver. The lesions observed in PSVD are microscopic (portal space and hepatic sinusoid).

1. How is PSVD diagnosed?

The diagnosis of PSVD requires histological analysis (under a microscope) of a liver tissue sample. This sample is obtained from a specimen taken under local anaesthesia, this procedure is known as liver biopsy. This examination enables to visualize abnormalities in the small vessels of the liver, which are only visible under the microscope. The liver biopsy also allows to rule-out cirrhosis, which is very important because the management of patients with PSVD differs from that of patients with cirrhosis.

Imaging tests such as Fibroscan (an ultrasound-like test that measures liver stiffness) or abdominal CT scan may show signs suggestive of PSVD, prompting the doctor to perform a liver biopsy.

Analysis of a liver tissue sample taken during a biopsy is mandatory for diagnosing PSVD.

2. What are the manifestations of PSVD?

a. Portal hypertension

Portal hypertension is the most common complication of PSVD (Figure 2). Portal hypertension occurs when the passage of blood through the liver is obstructed by damage to the small vessels of the liver. The main manifestations of portal hypertension in patients with PSVD are varices, splenomegaly, thrombocytopenia and ascites.

Varices are normal veins that have increased in size due to increased blood pressure. The most common varices are located in the esophagus and stomach (see separate insert).

Splenomegaly is an increase in the size of the spleen. In the vast majority of the patients splenomegaly is diagnosed at imaging (ultrasound or abdominal scan). Splenomegaly can cause discomfort or heaviness.

Thrombocytopenia is a decrease in the concentration of platelets in a blood sample. Thrombocytopenia does not increase the risk of bleeding from ruptured oesophageal varices. Severe thrombocytopenia can cause prolonged or heavy bleeding, such as menstrual bleeding.

Ascites is the accumulation of fluid in the abdominal cavity. Ascites is rare in people with PSVD and is usually transient, especially after a complication (such as bleeding). In exceptional cases, ascites is difficult to treat or is associated with chronic liver failure (liver dysfunction). These conditions may require liver transplantation.

a. Portal vein thrombosis

Portal vein thrombosis is the obstruction of the portal vein by a blood clot (Figure 2). Portal vein thrombosis may be favored by a condition associated with PSVD (such as a blood disorder that increases the risk of thrombosis). If portal vein thrombosis occurs, anticoagulant

treatment is started. Imaging (ultrasound or abdominal scan) is repeated every 6 months to screen for portal vein thrombosis.

b. Liver tests abnormalities

Some people have PSVD without portal hypertension. In this case, the only signs are mild abnormalities in liver blood tests, without any symptoms. Portal hypertension may appear after the diagnosis of PSVD. For this reason, medical follow-up is necessary.

Unlike cirrhosis, the risk of liver cancer is not increased in people with PVSD.

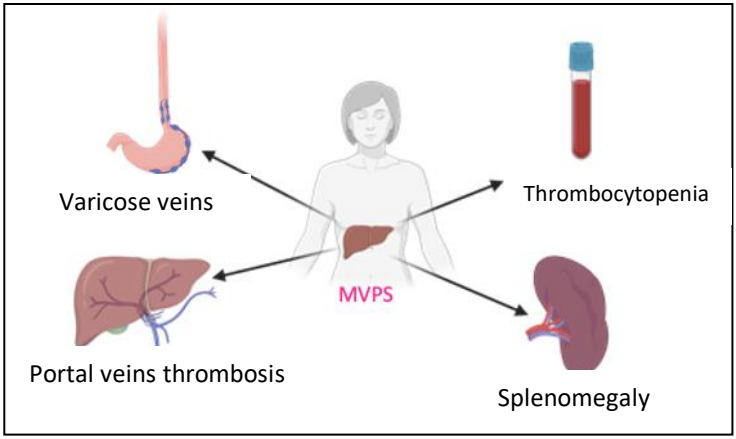

Figure 2: Manifestations of porto-sinusoidal vascular disease

3. How are people with MVPS treated?

a. Management of portal hypertension complications

Large varices can cause gastrointestinal bleeding (variceal rupture). Such bleeding or its recurrence can be largely prevented by medication (beta-blockers) or endoscopic treatment. Rarely, if the bleeding recurs and becomes disabling, radical treatment of portal hypertension (TIPS) may be proposed. The decision to undergo this type of procedure requires careful analysis in a specialized center (see separate leaflet).

Upper gastrointestinal endoscopy s performed when PSVD is diagnosed, and repeated periodically (every 1 to 3 years) to detect gastroesophageal varices. Recent studies suggest that when spleen stiffness (measurement of the spleen stiffness using Fibroscan) together with normal bilirubin concentration allows to rule out large gastroesophageal varices, and therefore, to avoid upper gastro intestinal endoscopy.

Specific treatments for thrombocytopenia and splenomegaly have not been proven to be effective and may be dangerous.

b. Workup and treatment of associated diseases

In about half of the people with PSVD, an associated predisposing factor is identified, usually after discussion with your doctor and analysis of blood tests. Factors associated with PSVD include certain medications, a prothrombotic state, haematological (blood) disorders, immune system disorders, or genetic disorders.

If an associated extrahepatic disease is diagnosed, a special treatment may be required. In this case, follow-up care is also organized by a doctor from another specialty (haematologist, internal medicine, etc.).

A systematic work-up is carried out to identify any factors that may contribute to the development of PSVD.

c. Anticoagulants

Anticoagulant therapy may be prescribed in a person with PSVD for several reasons: if a prothrombotic condition (i.e. a condition that increases the risk of thrombosis) is present, or if portal vein thrombosis is diagnosed. In addition, anticoagulant therapy may even be effective in preventing complications of PSVD (in particular portal vein thrombosis and complications of portal hypertension). This treatment is currently being evaluated in a French clinical trial (APIS trial, coordinated by Prof. Pierre-Emmanuel Rautou).

Anticoagulants are prescribed in cases of prothrombotic state or portal vein thrombosis.

In conclusion, PSVD is a rare disease involving lesions of the small hepatic vessels. Liver biopsy is mandatory to confirm the diagnosis. The main symptoms to look out for are gastroesophageal varices and portal vein thrombosis. Anticoagulant therapy may be prescribed for portal vein thrombosis, but may also be useful in preventing the complications of portal hypertension

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