Gastroenterol Hepatol. 2016;39(6):403–410

Gastroenterología y Hepatología

www.elsevier.es/gastroenterologia

REVIEW

# Treatment of non-cirrhotic, non-tumoural portal vein thrombosis☆

**Elba Llop**[a], **Susana Seijo**[b,*]

[a] *Hospital Universitario Puerta de Hierro Majadahonda, Madrid, Spain*
[b] *CTO, Department of Medicine, Icahn School of Medicine at Mount Sinai, New York, United States*

Received 17 June 2015; accepted 4 September 2015
Available online 26 May 2016

**KEYWORDS**
Portal hypertension;
Variceal gastrointestinal bleeding;
Anticoagulation;
Myeloproliferative neoplasia;
*JAK2V617F* mutation

**Abstract** Thrombosis of the splenoportal axis not associated with liver cirrhosis or neoplasms is a rare disease whose prevalence ranges from 0.7 to 3.7 per 100,000 inhabitants. However, this entity is the second most common cause of portal hypertension. Prothrombotic factors are present as an underlying cause in up to 70% of patients and local factors in 10–50%. The coexistence of several etiological factors is frequent. Clinical presentation may be acute or chronic (portal cavernomatosis). The acute phase can present as abdominal pain, nausea, vomiting, fever, rectorrhagia, intestinal congestion, and ischaemia. In this phase, early initiation of anticoagulation is essential to achieve portal vein recanalization and thus improve patient prognosis. In the chronic phase, symptoms are due to portal hypertension syndrome. In this phase, the aim of treatment is to treat or prevent the complications of portal hypertension. Anticoagulation is reserved to patients with a proven underlying thrombophilic factor.


**PALABRAS CLAVE**
Hipertensión portal;
Hemorragia digestiva varicosa;
Anticoagulación;
Neoplasia mieloproliferativa;
Mutación *JAK2V617F*

## Actuación ante la trombosis portal no cirrótica no tumoral

**Resumen** La trombosis del eje esplenoportal (TVP) no asociada a cirrosis hepática o neoplasias es una enfermedad rara con prevalencia que oscila entre el 0,7 y el 3,7 por 100.000 habitantes. Sin embargo, es la segunda causa de hipertensión portal. Hasta el 70% de los pacientes presentan factores protrombóticos como causa subyacente y entre el 10 y el 50%, factores locales. Es frecuente la coexistencia de varias entidades etiológicas. La presentación clínica puede ser aguda o crónica (cavernomatosis portal). La fase aguda se puede manifestar como dolor abdominal, náuseas, vómitos, fiebre, rectorragia, congestión intestinal e isquemia. Es esencial el inicio precoz de la anticoagulación en esta fase para conseguir la recanalización portal y, con ello,

☆ Please cite this article as: Llop E, Seijo S. Actuación ante la trombosis portal no cirrótica no tumoral. Gastroenterol Hepatol. 2016;39:403–410.

* Corresponding author.
*E-mail address:* susana.seijo@mssm.edu (S. Seijo).

2444-3824/

mejorar el pronóstico del paciente. En la fase de cavernomatosis portal, los síntomas vienen derivados del síndrome de hipertensión portal. En esta fase el tratamiento va dirigido a tratar o prevenir las complicaciones de la hipertensión portal. La anticoagulación quedará reservada a aquellos pacientes en los que se demuestre un factor trombofílico subyacente.


## Introduction

Portal vein thrombosis (PVT) is obstruction of the portal vein, with or without extension to other segments of the splanchnic venous system (splenic vein or superior mesenteric vein). The term, however, does not include isolated thrombosis of the splenic or superior mesenteric veins. Since PVT associated with cirrhosis or due to tumour invasion or compression has different therapeutic and prognostic implications, these should be considered different entities.[1–8] This paper refers only to non-cirrhotic, non-tumour PVT.

PVT can be classified as acute or chronic, constituting successive stages of the same disease. Although the causes are the same, clinical manifestations and management differ for each stage.[3–6,8–11]

Optimal management of patients with PVT is based initially on proper patient identification. Familiarity with the clinical manifestations of the disease will increase clinical suspicion, and evaluation of imaging tests by radiologists will be essential for making the correct diagnosis. Diagnosing the disease or risk factor that caused the thrombosis is also crucial. Finally, specific treatment of the PVT and its complications is vital.

## Clinical manifestations

The clinical manifestations depend on the developmental stage at which the PVT is detected (acute vs chronic) and the thrombus extension. PVT can be diagnosed in the acute phase, but this initial episode often goes unnoticed and the PVT is diagnosed in the chronic phase, when the patient has developed portal cavernoma. In recent years—thanks to better understanding of this disease and the availability and improvement of radiological techniques—this trend appears to be changing so that, at present, PVT is more frequently diagnosed in the acute phase. It is sometimes very difficult to differentiate an acute thrombosis from a re-thrombosis on a previously undetected portal cavernoma.

### Acute portal vein thrombosis

The main manifestation is usually abdominal pain, accompanied by non-specific symptoms such as fever, general malaise, systemic inflammatory response and dyspeptic symptoms (nausea and postprandial fullness). Symptoms often overlap with those of the local triggering factor (e.g. recent surgery, acute/chronic pancreatitis, colitis, etc.). If the thrombosis affects the mesenteric venous arches, the patient can develop intestinal ischaemia that can lead to intestinal necrosis. This should be suspected when there is abdominal pain, haematochezia, signs of peritonism, intra-abdominal free fluid and metabolic acidosis with renal or respiratory failure. The onset of intestinal stenosis may be the late sequela of mesenteric venous ischaemia. Recent series have shown a marked decrease in the incidence of intestinal ischaemia in patients in whom anticoagulation is instigated promptly.[2,11,14,15,17,18]

Gastro-oesophageal varices can appear early on (1 month after the acute episode), but in other patients, onset can be delayed a few months and then manifest in the form of variceal bleeding.[2,3,5,9,11] Early screening of varices 2–3 months after the acute episode is therefore recommended in patients with acute PVT, and again at 6–9 months in cases in which the varices are not present in the initial endoscopy and the PVT has not been recanalized.[2,8,11–17]

Isolated thrombosis of a large intrahepatic portal vein branch (lobar or segmental) is a special situation that is often detected incidentally,[5,9,11,14,15] and can be accompanied by a moderate, transient increase in transaminases. Atrophy of the affected hepatic territory may subsequently occur, with hypertrophy of the rest of the parenchyma, but portal hypertension does not usually develop.

### Chronic portal vein thrombosis. Portal cavernoma

After an acute PVT episode, the collaterals that make up the cavernoma form within a few days and stabilise within 3–5 weeks.[12,15,19] At the same time, vasodilation of the hepatic arterial territory occurs in response to decreased portal venous flow.[14,18] The activation of both mechanisms enables the total hepatic blood flow to remain stable or decrease only minimally, while portal vein pressure increases.

Portal cavernoma or chronic PVT is usually diagnosed incidentally during an endoscopic or radiological study performed in a patient with thrombocytopenia, splenomegaly or other signs of portal hypertension.[3–6,8,10,14,20] The complications of portal hypertension are the main clinical manifestations of portal cavernoma. Early endoscopy is therefore recommended to rule out the presence of varices. There is no evidence to recommend a specific screening regimen for varices in patients in whom these are not observed in the initial endoscopy. However, once the cavernoma has been established and ''stabilised'', the risk of varices in patients who have not already developed them in the early years might be lower. Nevertheless, further research is needed to confirm this, so current recommendations suggest