Chronic portal vein thrombosis in adults: Clinical manifestations, diagnosis, and management

AUTHOR: Arun J Sanyal, MD
SECTION EDITOR: Sanjiv Chopra, MD, MACP
DEPUTY EDITOR: Kristen M Robson, MD, MBA, FACG

All topics are updated as new evidence becomes available and our peer review process is complete.

Literature review current through: **Sep 2023.**
This topic last updated: **Jan 30, 2023.**

INTRODUCTION

The portal vein is formed by the confluence of the splenic and superior mesenteric veins, which drain the spleen and small intestine, respectively (figure 1). Occlusion of the portal vein by thrombus (portal vein thrombosis [PVT]) typically occurs in patients with cirrhosis and/or prothrombotic disorders (table 1). Chronic PVT develops in patients with acute PVT that does not resolve (with or without treatment). Patients with chronic PVT develop collateral blood vessels that bring blood in a hepatopetal manner around the area of obstruction, known as cavernous transformation of the portal vein or portal cavernoma. When seen in a transverse section, as on a computed tomographic (CT) scan, cavernous transformation gives the appearance of multiple caveolar orifices (image 1). Complications of chronic PVT include portal hypertension and portal cholangiopathy.

This topic will review the clinical manifestations, diagnosis, and management of chronic PVT. The epidemiology and pathogenesis of PVT, the approach to patients with acute PVT, and other causes of noncirrhotic portal hypertension are discussed elsewhere. (See "Epidemiology and pathogenesis of portal vein thrombosis in adults" and "Acute portal vein thrombosis in adults: Clinical manifestations, diagnosis, and management" and "Noncirrhotic portal hypertension".)

The approach to patients with PVT has also been reviewed in society guidelines [1,2]. The discussion that follows is generally consistent with those guidelines.

CLINICAL MANIFESTATIONS

Chronic portal vein thrombosis (PVT) may be asymptomatic and discovered incidentally when abdominal imaging is obtained for other reasons, or patients may present with symptoms related to portal hypertension or portal cholangiopathy, two of the complications of chronic PVT. Patients with chronic PVT may also develop intestinal ischemia and infarction if there is extension of the clot into the superior mesenteric vein, although the risk is small [3]. Patients with chronic PVT may also have clinical manifestations related underlying conditions that predisposed to PVT, such as cirrhosis (table 1). (See "Epidemiology and pathogenesis of portal vein thrombosis in adults", section on 'Portal hypertension' and "Epidemiology and pathogenesis of portal vein thrombosis in adults", section on 'Portal cholangiopathy' and "Cirrhosis in adults: Etiologies, clinical manifestations, and diagnosis", section on 'Clinical manifestations'.)

Patients with chronic PVT, even if asymptomatic, frequently have esophageal or gastric varices, and the most common clinical presentation is gastrointestinal bleeding [4-7]. In a retrospective series that included 40 patients with chronic PVT, at the time of diagnosis, esophageal varices were present in 35 patients (88 percent), gastric varices in 20 patients (50 percent), portal hypertensive gastropathy in 19 patients (48 percent), and gastrointestinal bleeding in 19 patients (48 percent) [4]. The association of chronic PVT with variceal bleeding probably reflects that the clinical presentation of gastrointestinal bleeding leads to the diagnosis of PVT.

Portal cholangiopathy (also referred to as portal biliopathy) is also common in patients with longstanding chronic PVT and is due to compression of the large bile ducts by the venous collaterals that form in patients with chronic PVT. Studies using magnetic resonance cholangiopancreatography have found evidence of portal cholangiopathy in the majority of patients with long-standing chronic PVT [8,9]. (See "Epidemiology and pathogenesis of portal vein thrombosis in adults", section on 'Portal cholangiopathy'.)

Most patients with portal cholangiopathy are asymptomatic, though some develop biliary complications including pruritus, obstructive jaundice, cholecystitis, and cholangitis [8,10-15]. As an example, in a series of 21 patients, only three (14 percent) developed symptoms [10]. Two had obstructive jaundice, and one had cholangitis. In a second series, 5 of 23 patients were found to have choledocholithiasis, but all were asymptomatic at the time of diagnosis [15]. Four of the patients were followed, and three subsequently developed cholangitis.

Symptoms — Patients with chronic PVT may be asymptomatic from the thrombosis, particularly if they have underlying cirrhosis [3]. Among patients who are symptomatic, gastrointestinal

bleeding is the most common clinical presentation. Patients with gastrointestinal bleeding typically report hematemesis, melena, or hematochezia [16]. Patients may also have symptoms related to portal cholangiopathy or intestinal ischemia. Symptoms seen with portal cholangiopathy include jaundice, pruritus, or biliary colic; the presence of jaundice, abdominal pain, and fever is suggestive of cholangitis. If intestinal ischemia or infarction develops, patients may report abdominal pain that radiates to the back, abdominal distension from ascites, or bloody diarrhea. (See "Acute cholangitis: Clinical manifestations, diagnosis, and management", section on 'Clinical manifestations' and "Mesenteric venous thrombosis in adults", section on 'Clinical presentations' and "Nonocclusive mesenteric ischemia", section on 'Clinical features'.)

Patients may also have symptoms related to cirrhosis or other conditions that predispose to the development of PVT (table 1). (See "Cirrhosis in adults: Etiologies, clinical manifestations, and diagnosis", section on 'Clinical manifestations'.)

Physical examination — Physical examination in patients with chronic PVT may be normal, though it frequently is notable for signs of portal hypertension, such as splenomegaly. Patients may also have findings suggestive of underlying cirrhosis, such as palmar erythema or a fluid wave from ascites. (See "Cirrhosis in adults: Etiologies, clinical manifestations, and diagnosis", section on 'Physical examination'.)

Splenomegaly has been reported in 25 to 100 percent of patients with chronic PVT [6,17,18]. The mechanism behind splenomegaly is not fully understood, but it may be related to increased venous congestion and splenic arterial flow. The degree of splenomegaly in patients with chronic PVT without cirrhosis is often greater than is seen in those with cirrhosis, and massive splenomegaly may be present [19].

A small amount of ascites is seen in 10 to 20 percent of patients with chronic PVT who do not have cirrhosis [6,17], particularly after gastrointestinal bleeding with massive fluid resuscitation, which may lead to acute dilutional hypoalbuminemia. (See "Evaluation of adults with ascites".)

Subclinical encephalopathy is seen in half of patients with portal vein thrombosis without cirrhosis, though overt encephalopathy is uncommon and is typically seen following gastrointestinal bleeding, if there is renal failure, or in the setting of sepsis in older adults [20,21]. (See "Hepatic encephalopathy in adults: Clinical manifestations and diagnosis", section on 'Clinical manifestations'.)

Laboratory testing — Liver biochemical tests, such as the serum aminotransferases, are usually normal or only slightly increased. In the absence of cirrhosis, measures of hepatic synthetic function are typically preserved with the exception of hypoalbuminemia, which may be seen after fluid resuscitation for gastrointestinal bleeding. Liver failure should not occur

unless patients have concomitant cirrhosis [7]. Patients may also have findings associated with hypersplenism such as anemia, thrombocytopenia, and leukopenia. In patients with portal cholangiopathy, laboratory testing may reveal a cholestatic pattern of laboratory test abnormalities (eg, elevated alkaline phosphatase and bilirubin). (See "Tests of the liver's biosynthetic capacity (eg, albumin, coagulation factors, prothrombin time)" and "Enzymatic measures of cholestasis (eg, alkaline phosphatase, 5'-nucleotidase, gamma-glutamyl transpeptidase)".)

Abdominal imaging — Radiographic findings in chronic PVT include demonstration of cavernous transformation of the portal vein as well as filling defects within the portal vein. (See 'Diagnosis' below.)

DIAGNOSIS

Chronic portal vein thrombosis (PVT) is diagnosed with abdominal imaging. Our approach is to start with abdominal ultrasound with Doppler imaging, followed by a contrast-enhanced computed tomographic (CT) scan or magnetic resonance imaging (MRI) to confirm the diagnosis and to look for predisposing conditions, such as hepatocellular carcinoma. We start with Doppler ultrasound because it is inexpensive and can identify biliary pathology that may lead to abdominal pain. It is reasonable to start with a contrast-enhanced CT or MRI if suspicion for chronic PVT is high or if there is generalized abdominal pain, in which case the differential diagnosis is large.

Patients should also undergo a serologic evaluation for potential predisposing conditions. While it is our practice to do a hypercoagulability workup in all patients with PVT, including those with decompensated cirrhosis, others would not perform the evaluation in patients with decompensated cirrhosis due to rebalanced coagulation pathways, which can tilt from a hypercoagulable to hypocoagulable state and which are not due to a pre-existing condition. (See "Epidemiology and pathogenesis of portal vein thrombosis in adults", section on 'Pathogenesis'.)

Abdominal ultrasound — Abdominal ultrasound with Doppler imaging in a patient with chronic PVT demonstrates hyperechoic material within the portal vein that may extend into the mesenteric or splenic veins, dilation of the portal vein and its tributaries, and absence of flow within the portal vein [22]. Ultrasonography may also reveal a mass of tortuous vessels at the porta hepatis or within the liver [23]. The velocity of blood flow within those vessels is usually less than that in a normal portal vein [24].

Abdominal CT — An abdominal CT scan in a patient with chronic PVT reveals a network of intertwined, densely packed veins in the hepatoduodenal ligament and porta hepatis. The thrombosed portal segment is usually not seen, but small veins may show enhancement [25]. In addition, communication between collateral vessels and intrahepatic portal veins may be seen [26].

Abdominal MRI — Findings on abdominal MRI in a patient with PVT include portal vein occlusion as well as collateral veins around the porta hepatis. On MRI angiography, PVT appears as a filing defect that partially or completely occludes the vessel lumen in the portal venous phase [27]. In one study, MRI had a sensitivity of 100 percent and a specificity of 99 percent for detecting PVT [28]. MRI is sensitive for detecting submucosal, serosal, and periesophageal collaterals and may demonstrate portal flow reduction or inversion [29].

Angiography — If the diagnosis of chronic PVT is in doubt after obtaining standard radiographic imaging, or if shunt surgery is planned, angiography may be considered. During contrast injection, PVT is usually seen as a filling defect or non-opacification of the portal vein or one of its branches. With cavernous transformation, venous branches are often filled by the collateral veins (image 2) [23]. Angiography is rarely used due to increasing utilization of CT scan and MRI.

Identification of predisposing conditions — Patients with PVT who do not have cirrhosis or who have compensated (Child-Pugh class A or B) cirrhosis should be evaluated for conditions that may have predisposed to thrombosis, such as hypercoagulable states. In those with decompensated cirrhosis, the decision to pursue a workup for a genetic basis for hypercoagulability should be individualized. We often reserve it for recurrent thrombotic events or if thrombosis occurs in more than one vascular bed or if there is a family history of thrombotic disorders. The approach to the evaluation of patients with established venous thrombosis is discussed in detail elsewhere. (See "Evaluating adult patients with established venous thromboembolism for acquired and inherited risk factors".)

DIFFERENTIAL DIAGNOSIS

The primary considerations in the differential diagnosis of chronic portal vein thrombosis (PVT) are invasion of the portal vein by an abdominal malignancy (most frequently hepatocellular carcinoma) or, less often, constriction of the portal vein within a tumor (typically pancreatic cancer or cholangiocarcinoma) [30]. In addition, portal cavernomas may resemble cholangiocarcinoma or a pancreatic head mass on imaging studies. Endoscopic ultrasound and magnetic resonance angiography can be used to differentiate chronic PVT from

cholangiocarcinoma or pancreatic cancer [31]. (See "Acute portal vein thrombosis in adults: Clinical manifestations, diagnosis, and management", section on 'Differential diagnosis' and "Clinical manifestations, diagnosis, and staging of exocrine pancreatic cancer", section on 'Diagnostic approach' and "Clinical manifestations and diagnosis of cholangiocarcinoma", section on 'Imaging studies'.)

In cases of tumor invasion or external constriction, a thrombus within the portal vein may develop as a secondary event (malignant PVT). Imaging characteristics can help differentiate benign PVT from malignant PVT. Differentiating benign from malignant PVT is particularly important in patients with cirrhosis and hepatocellular carcinoma who are being considered for liver transplantation since malignant PVT is a contraindication to liver transplantation.

Findings that suggest a malignant PVT include [32]:

- Elevated alpha fetoprotein
- Portal vein diameter >23 mm
- Enhancement of endoluminal material during the arterial phase of contrast injection
- Arterial-like pulsatile flow seen with Doppler ultrasound
- Disruption of the vessel walls

MANAGEMENT

The management of chronic portal vein thrombosis (PVT) depends on the presence of predisposing conditions and the patient's comorbidities. Basic management includes screening for esophageal varices and treating complications of portal hypertension and portal cholangiopathy. In addition, anticoagulation may be indicated for some patients.

- (See "Methods to achieve hemostasis in patients with acute variceal hemorrhage".)
- (See "Ascites in adults with cirrhosis: Initial therapy".)
- (See "Hepatic encephalopathy in adults: Treatment".)
- (See "Pruritus associated with cholestasis", section on 'Management'.)
- (See "Acute cholangitis: Clinical manifestations, diagnosis, and management", section on 'Management'.)

Screening for varices — Patients with chronic PVT should be screened for esophageal varices [1]. While not specifically tested in patients with varices due to PVT, it is reasonable to administer nonselective beta blockers or endoscopic therapy to decrease the risk of variceal bleeding in those found to have varices. (See "Primary prevention of bleeding from esophageal

varices in patients with cirrhosis" and "Prevention of recurrent bleeding from esophageal varices in patients with cirrhosis".)

Anticoagulation

Goals of therapy — The goal of anticoagulation is to prevent recurrent thrombosis, prevent thrombus extension, maintain splanchnic venous drainage, relieve symptoms, and promote recanalization in patients with cirrhosis. However, while patients with chronic PVT are often at risk for recurrent thrombosis, they are also at risk for bleeding. As a result, the decision to start anticoagulation must be made on a case-by-case basis.

Selecting patients for treatment — We base the decision to anticoagulate on the following:

- What is the patient's risk of bleeding (eg, does the patient have large varices with features associated with an increased risk of bleeding)? (See "Pathogenesis of variceal bleeding in patients with cirrhosis", section on 'Predictive factors'.)
- What is the patient's risk of a thrombotic event (eg, does the patient have an underlying prothrombotic disorder)? (See "Epidemiology and pathogenesis of portal vein thrombosis in adults", section on 'Pathogenesis'.)
- How likely is the patient to survive a bleed or a thrombotic event?
- Is the patient awaiting liver transplantation?
- Does the patient have symptoms (eg, abdominal pain)?

In general, we offer anticoagulation to patients who are at increased risk for recurrent thrombosis based on their clinical history or laboratory studies. In such patients, we suggest long-term anticoagulation and discuss the risk of adverse events (eg, bleeding). We give anticoagulation to patients with a history of gastrointestinal variceal bleeding or large varices who are at increased risk for bleeding (particularly in those with cirrhosis) only if adequate measures to prevent recurrent bleeding can be implemented. We prefer nonselective beta blockers over variceal ligation for prophylaxis in such patients because of the potential risk of bleeding from esophageal ulcers that form when the bands used for variceal ligation slough off. (See "Prevention of recurrent bleeding from esophageal varices in patients with cirrhosis" and "Primary prevention of bleeding from esophageal varices in patients with cirrhosis", section on 'Preventive strategies' and "Epidemiology and pathogenesis of portal vein thrombosis in adults", section on 'Pathogenesis'.)

We also offer anticoagulation to patients with PVT and advanced cirrhosis who are awaiting transplantation and do not have contraindications to anticoagulation.

Choice of agent — When treating with anticoagulation, we often use enoxaparin rather than warfarin because of its shorter duration of action, less variability in anticoagulation, decreased need for monitoring, and decreased difficulty when managing patients around the time of liver transplantation. An alternative is to use an oral anticoagulant (if warfarin is used, our goal international normalized ratio [INR] is 2 to 3) [33].

Direct oral anticoagulant (DOAC) therapy is an alternative to enoxaparin or warfarin for treating chronic PVT. DOAC use is individualized and informed by the potential benefits (eg, oral administration, decreased need for laboratory monitoring), the risk of bleeding, and patient preferences. Data on efficacy and safety of DOACs for PVT have been mixed [34-38]. In a cohort study including 40 patients with chronic PVT and cirrhosis, DOAC therapy (rivaroxaban or dabigatran) for three or six months was associated with partial/complete recanalization rates of 13 and 28 percent, respectively [34]. The risk of bleeding was not significantly higher for patients on DOACs compared with a historic control group. In another study including 50 patients with PVT and cirrhosis who were initially treated with danaparoid, subsequent therapy with edoxaban was associated with reduced volume of thrombus compared with warfarin, although the clinical implications of this outcome were uncertain [35]. Bleeding rates were numerically higher in the edoxaban group (15 versus 7 percent). In a report of three patients with PVT and cirrhosis, dabigatran was not associated with recanalization in any of the patients [37].

For patients with chronic PVT in the absence of cirrhosis, data comparing DOACs with other anticoagulants have been limited to observational studies. In a study including 63 patients without cirrhosis who had PVT, DOAC therapy was associated with higher rates of complete PVT resolution on imaging compared with warfarin (96 versus 55 percent) [36].

Efficacy

Patients without cirrhosis — Studies have suggested that anticoagulation for PVT in the absence of cirrhosis resulted in favorable outcomes [20,36,38]. In a trial including 111 patients without cirrhosis who had chronic PVT, rivaroxaban resulted in lower risk of recurrent thrombosis compared with no anticoagulation after a median follow up of 11.8 months (0 versus 19.7 events per 100 person-years) [38]. In addition, the risk of bleeding was not significantly different between groups. Although DOACs were not associated with an increased risk of bleeding, there remains a theoretical risk that we discuss with the patient when initiating anticoagulation. (See 'Choice of agent' above.)

Patients with cirrhosis — Observational data have suggested that anticoagulation may be beneficial for patients with cirrhosis and portal vein thrombosis. In a meta-analysis of eight studies including 353 patients with cirrhosis and PVT, patients treated with anticoagulants (ie, low weight heparin or warfarin) had higher rates of either partial or complete recanalization compared with untreated patients (71 versus 42 percent; OR 4.8, 95% CI 2.7-8.7) [39]. The overall rate of bleeding (ie, both minor and major episodes) was similar in the anticoagulated and untreated patients (11 percent in both groups). The risk of variceal bleeding was assessed in four studies including 158 patients, and the variceal bleeding rate was lower in anticoagulated patients compared with untreated patients (2 versus 12 percent; OR 0.23, 95% CI 0.06-0.94). A clinical trial is needed to validate the efficacy and safety of anticoagulant therapy in this setting and identify which patients should be treated.

Data on the use of anticoagulation in patients with cirrhosis awaiting liver transplantation are limited. In a study of 19 patients with cirrhosis who were awaiting liver transplantation, anticoagulation was associated with restoration of portal vein patency in 10, compared with 0 of 10 historic controls [40]. Anticoagulation also reduced postoperative complications, especially in those with extensive clot.

Portal cholangiopathy — Data are limited regarding the treatment of portal cholangiopathy. Patients with jaundice or recurrent biliary symptoms due to portal cholangiopathy may benefit from endoscopic removal of bile duct stones followed by insertion of a biliary stent or creation of a surgical portal-systemic shunt. In a series of six patients, good outcomes were achieved in two patients with stenting [41]. Four patients failed attempts at endoscopic treatment but did well after surgical portal-systemic shunting. Biliary decompression should be performed prior to biliary surgery because instrumentation of an obstructed biliary tract puts patients at high risk for cholangitis. (See "Acute cholangitis: Clinical manifestations, diagnosis, and management", section on 'Pathogenesis'.)

Management of complications — Complications of chronic PVT include variceal bleeding, ascites, hepatic encephalopathy, pruritus, and cholangitis. Patients with these complications are managed similarly to those who have these complications without PVT. (See "Methods to achieve hemostasis in patients with acute variceal hemorrhage" and "Ascites in adults with cirrhosis: Initial therapy" and "Hepatic encephalopathy in adults: Treatment" and "Pruritus associated with cholestasis", section on 'Management'.)

Patients with variceal hemorrhage are treated with endoscopic therapy, though patients with repeated variceal hemorrhage may need additional therapy. Definitive salvage therapy should be considered for patients who experience repeated hemorrhage despite adequate endoscopic treatment, have isolated varices in the gastric fundus, or have ectopic varices (eg, splenectomy

or surgical shunting depending upon the site of bleeding). Surgery corrects the varices and is usually well-tolerated because liver function typically remains well-preserved in the absence of cirrhosis. Recurrent hemorrhage due to portal hypertension related to extensive portal vein thrombosis is clinically uncommon and if a collateral vessel is large enough to place a transjugular intrahepatic portosystemic shunt (TIPS) is present, an attempt to decompress the portal system by a TIPS should be made. However, there is only sparse data related to the efficacy of this intervention. Experience with the Suguira procedure in Western countries is limited and should be attempted only at centers with expertise. The severity of underlying liver failure also dictates the surgical risk.

The choice of surgery depends upon the anatomy of obstruction:

- Splenectomy is used for patients with splenic vein thrombosis and bleeding gastric varices (figure 1) [42].

- In patients with diffuse thrombosis of the portal, mesenteric, and splenic veins, a shunt operation may not be possible. Such patients may benefit from a non-shunting operation, such as a modified Sugiura procedure in which the esophagus is transected while the paraesophagogastric region is devascularized, though this procedure is rarely performed [43,44]. (See "Methods to achieve hemostasis in patients with acute variceal hemorrhage", section on 'Surgery'.)

- In patients in whom the superior mesenteric vein is patent, a mesocaval shunt combined with splenectomy and left gastric vein ligature can be performed. Some patients may have a large spontaneous splenorenal collateral, which must be tied off during the procedure.

Transjugular intrahepatic portosystemic shunting (TIPS) (figure 2) has been shown to be technically feasible in some cases of extrahepatic portal vein thrombosis, such as patients without cavernous transformation in whom the thrombosed vein can be accessed, dilated, and stented, and it may be considered in selected cases with symptoms related to portal hypertension that fail to respond to other treatments [45-47]. However, the ability to adequately decompress the portal vein is unpredictable. In addition, although TIPS may be technically feasible in patients with cavernous transformation, it is unlikely to adequately decompress the liver in all cases [48,49]. When performed, it is advisable to perform a thorough assessment for prothrombotic states and consider anticoagulation after the bleeding is controlled. (See "Overview of transjugular intrahepatic portosystemic shunts (TIPS)".)

In patients with impending bowel ischemia due to PVT, the decision regarding the best approach to decompress the portal vein and preserve the bowel (eg, surgical shunting or TIPS)

should be individualized based on the patient's comorbidities, technical feasibility, and available expertise. (See "Mesenteric venous thrombosis in adults", section on 'Anticoagulation'.)

PREVENTION IN PATIENTS WITH CIRRHOSIS

Prevention of PVT in individuals with cirrhosis focuses on optimizing hepatic function, reducing portal venous pressure, and increasing portal flow, which diminishes stasis. These issues are presented separately. (See "Hemostatic abnormalities in patients with liver disease", section on 'Portal vein thrombosis (PVT)'.)

PROGNOSIS

Patients who receive treatment for chronic PVT and its complications have a good prognosis in the absence of cirrhosis or malignancy. In a study of 136 patients with chronic PVT who did not have cirrhosis or an underlying malignancy, fewer than 5 percent of patients followed for five years died from PVT complications, such as intestinal infarction or gastrointestinal bleeding [20], though mortality may be higher for those with PVT that also involves the mesenteric vein [50]. Mortality is more often related to age, the cause of PVT (eg, myeloproliferative disorder or decompensated cirrhosis), or unrelated diseases.

SOCIETY GUIDELINE LINKS

Links to society and government-sponsored guidelines from selected countries and regions around the world are provided separately. (See "Society guideline links: Hepatic, portal, and splenic vein thrombosis".)

SUMMARY AND RECOMMENDATIONS

- **Clinical manifestations** – Chronic portal vein thrombosis (PVT) may be asymptomatic and discovered incidentally when abdominal imaging is obtained for other reasons, or patients may present with symptoms related to portal hypertension or portal cholangiopathy, two of the complications of chronic PVT. Patients with chronic PVT may also develop intestinal ischemia and infarction if there is extension of the clot into the superior mesenteric vein, though the risk is small. Patients with chronic PVT may also have clinical manifestations related to underlying conditions that predisposed to PVT, such as cirrhosis (table 1). (See 'Clinical manifestations' above.)

- **Diagnosis** – Chronic PVT is diagnosed with abdominal imaging. Our approach is to start with abdominal ultrasound with Doppler imaging, followed by a contrast-enhanced computed tomographic scan or magnetic resonance imaging to confirm the diagnosis and to look for predisposing conditions, such as hepatocellular carcinoma. (See 'Diagnosis' above.)

- **Evaluating for predisposing conditions** – Patients with PVT should be evaluated for conditions that may have predisposed to the development of the clot, such as prothrombotic states. While it is our practice to perform a hypercoagulability workup in all patients with PVT, including those with decompensated (Child C class) cirrhosis, others would not perform the evaluation in patients with decompensated cirrhosis because of the high prevalence of PVT in this population. (See 'Identification of predisposing conditions' above.)

- **Differential diagnosis** – The primary considerations in the differential diagnosis of chronic PVT are invasion of the portal vein by an abdominal malignancy (most frequently hepatocellular carcinoma) or, less often, constriction of the portal vein within a tumor (typically pancreatic cancer or cholangiocarcinoma). In addition, portal cavernomas may resemble cholangiocarcinoma or a pancreatic head mass on imaging studies. (See 'Differential diagnosis' above.)

- **Management** – The management of chronic PVT depends on the presence of predisposing conditions and the patient's comorbidities. General measures include screening for esophageal varices and treating complications of portal hypertension and portal cholangiopathy:

  - (See "Primary prevention of bleeding from esophageal varices in patients with cirrhosis", section on 'Preventive strategies'.)
  - (See "Methods to achieve hemostasis in patients with acute variceal hemorrhage".)
  - (See "Ascites in adults with cirrhosis: Initial therapy".)
  - (See "Hepatic encephalopathy in adults: Treatment".)
  - (See "Pruritus associated with cholestasis", section on 'Management'.)

  In addition, anticoagulation may be indicated for some patients.

  The goal of anticoagulation in patients with chronic PVT is to prevent recurrent thrombosis, prevent thrombus extension, maintain splanchnic venous drainage, relieve symptoms, and promote recanalization in patients with cirrhosis. However, while patients with chronic PVT are often at risk for recurrent thrombosis, they are also at risk for

bleeding. As a result, the decision to start anticoagulation must be made on a case-by-case basis. (See 'Anticoagulation' above.)

We base the decision to use anticoagulation on the following:

- What is the patient's risk of bleeding (eg, does the patient have large varices with features associated with an increased risk of bleeding)? (See "Pathogenesis of variceal bleeding in patients with cirrhosis", section on 'Predictive factors'.)
- What is the patient's risk of a thrombotic event (eg, does the patient have an underlying prothrombotic disorder)? (See "Epidemiology and pathogenesis of portal vein thrombosis in adults", section on 'Pathogenesis'.)
- How likely is the patient to survive a bleed or a thrombotic event?
- Is the patient awaiting liver transplantation?
- Does the patient have symptoms (eg, abdominal pain)?

For patients who are at increased risk for recurrent thrombosis based on their clinical history or laboratory studies or who are awaiting liver transplantation, we suggest anticoagulation rather than expectant management, provided they are not at increased risk for bleeding or death were bleeding to occur (**Grade 2C**). Patients at increased risk for thrombosis include those with inherited prothrombotic disorders, decompensated cirrhosis, or malignancy. (See 'Anticoagulation' above and "Epidemiology and pathogenesis of portal vein thrombosis in adults", section on 'Pathogenesis'.)

For patients who are at increased risk for bleeding, are unlikely to survive a bleeding episode, or are not at increased risk for recurrent thrombosis, we suggest expectant management rather than anticoagulation (**Grade 2C**). Patients at increased risk of bleeding include those with large varices who have not had adequate prophylactic measures to prevent bleeding. Patients not at increased risk for thrombosis include those with PVT that developed due to a transient event, such as transient hypovolemia, and who do not have any underlying disorders that predispose to thrombosis. (See 'Anticoagulation' above.)

REFERENCES

1. Northup PG, Garcia-Pagan JC, Garcia-Tsao G, et al. Vascular Liver Disorders, Portal Vein Thrombosis, and Procedural Bleeding in Patients With Liver Disease: 2020 Practice Guidance by the American Association for the Study of Liver Diseases. Hepatology 2021; 73:366.
2. Simonetto DA, Singal AK, Garcia-Tsao G, et al. ACG Clinical Guideline: Disorders of the Hepatic and Mesenteric Circulation. Am J Gastroenterol 2020; 115:18.
3. Hoekstra J, Janssen HL. Vascular liver disorders (II): portal vein thrombosis. Neth J Med 2009; 67:46.
4. Sogaard KK, Astrup LB, Vilstrup H, Gronbaek H. Portal vein thrombosis; risk factors, clinical presentation and treatment. BMC Gastroenterol 2007; 7:34.
5. Amitrano L, Guardascione MA, Brancaccio V, et al. Risk factors and clinical presentation of portal vein thrombosis in patients with liver cirrhosis. J Hepatol 2004; 40:736.
6. Webb LJ, Sherlock S. The aetiology, presentation and natural history of extra-hepatic portal venous obstruction. Q J Med 1979; 48:627.
7. Kameda H, Yamazaki K, Imai F, et al. Obliterative portal venopathy: A comparative study of 184 cases of extrahepatic portal obstruction and 469 cases of idiopathic portal hypertension. J Gastroenterol Hepatol 1986; 1:139.
8. Condat B, Vilgrain V, Asselah T, et al. Portal cavernoma-associated cholangiopathy: a clinical and MR cholangiography coupled with MR portography imaging study. Hepatology 2003; 37:1302.
9. Llop E, de Juan C, Seijo S, et al. Portal cholangiopathy: radiological classification and natural history. Gut 2011; 60:853.
10. Khuroo MS, Yattoo GN, Zargar SA, et al. Biliary abnormalities associated with extrahepatic portal venous obstruction. Hepatology 1993; 17:807.
11. Chattopadhyay S, Nundy S. Portal biliopathy. World J Gastroenterol 2012; 18:6177.
12. Malkan GH, Bhatia SJ, Bashir K, et al. Cholangiopathy associated with portal hypertension: diagnostic evaluation and clinical implications. Gastrointest Endosc 1999; 49:344.
13. Dilawari JB, Chawla YK. Pseudosclerosing cholangitis in extrahepatic portal venous obstruction. Gut 1992; 33:272.
14. Dhiman RK, Puri P, Chawla Y, et al. Biliary changes in extrahepatic portal venous obstruction: compression by collaterals or ischemic? Gastrointest Endosc 1999; 50:646.
15. Sarin SK, Bhatia V, Markwane U. Portal biliopathy in extra hepatic portal vein obstruction. Indian J Gastroenterol 1992; 2:A82.

16. Sharma B, Raina S, Sharma R. Bleeding ectopic varices as the first manifestation of portal hypertension. Case Reports Hepatol 2014; 2014:140959.
17. Handa P, Crowther M, Douketis JD. Portal vein thrombosis: a clinician-oriented and practical review. Clin Appl Thromb Hemost 2014; 20:498.
18. Cohen J, Edelman RR, Chopra S. Portal vein thrombosis: a review. Am J Med 1992; 92:173.
19. Blendis LM, Banks DC, Ramboer C, Williams R. Spleen blood flow and splanchnic haemodynamics in blood dyscrasia and other splenomegalies. Clin Sci 1970; 38:73.
20. Condat B, Pessione F, Hillaire S, et al. Current outcome of portal vein thrombosis in adults: risk and benefit of anticoagulant therapy. Gastroenterology 2001; 120:490.
21. Mínguez B, García-Pagán JC, Bosch J, et al. Noncirrhotic portal vein thrombosis exhibits neuropsychological and MR changes consistent with minimal hepatic encephalopathy. Hepatology 2006; 43:707.
22. Van Gansbeke D, Avni EF, Delcour C, et al. Sonographic features of portal vein thrombosis. AJR Am J Roentgenol 1985; 144:749.
23. Hidajat N, Stobbe H, Griesshaber V, et al. Imaging and radiological interventions of portal vein thrombosis. Acta Radiol 2005; 46:336.
24. De Gaetano AM, Lafortune M, Patriquin H, et al. Cavernous transformation of the portal vein: patterns of intrahepatic and splanchnic collateral circulation detected with Doppler sonography. AJR Am J Roentgenol 1995; 165:1151.
25. Mori H, Hayashi K, Uetani M, et al. High-attenuation recent thrombus of the portal vein: CT demonstration and clinical significance. Radiology 1987; 163:353.
26. Gabata T, Matsui O, Kadoya M, et al. Gallbladder varices: demonstration of direct communication to intrahepatic portal veins by color doppler sonography and CT during arterial portography. Abdom Imaging 1997; 22:82.
27. Catalano OA, Choy G, Zhu A, et al. Differentiation of malignant thrombus from bland thrombus of the portal vein in patients with hepatocellular carcinoma: application of diffusion-weighted MR imaging. Radiology 2010; 254:154.
28. Kreft B, Strunk H, Flacke S, et al. Detection of thrombosis in the portal venous system: comparison of contrast-enhanced MR angiography with intraarterial digital subtraction angiography. Radiology 2000; 216:86.
29. Squillaci E, Crecco M, Lo Presti G, et al. [Angiography with magnetic resonance in the study of portal hypertension. Clinical applications]. Radiol Med 1993; 85:756.
30. Primignani M. Portal vein thrombosis, revisited. Dig Liver Dis 2010; 42:163.

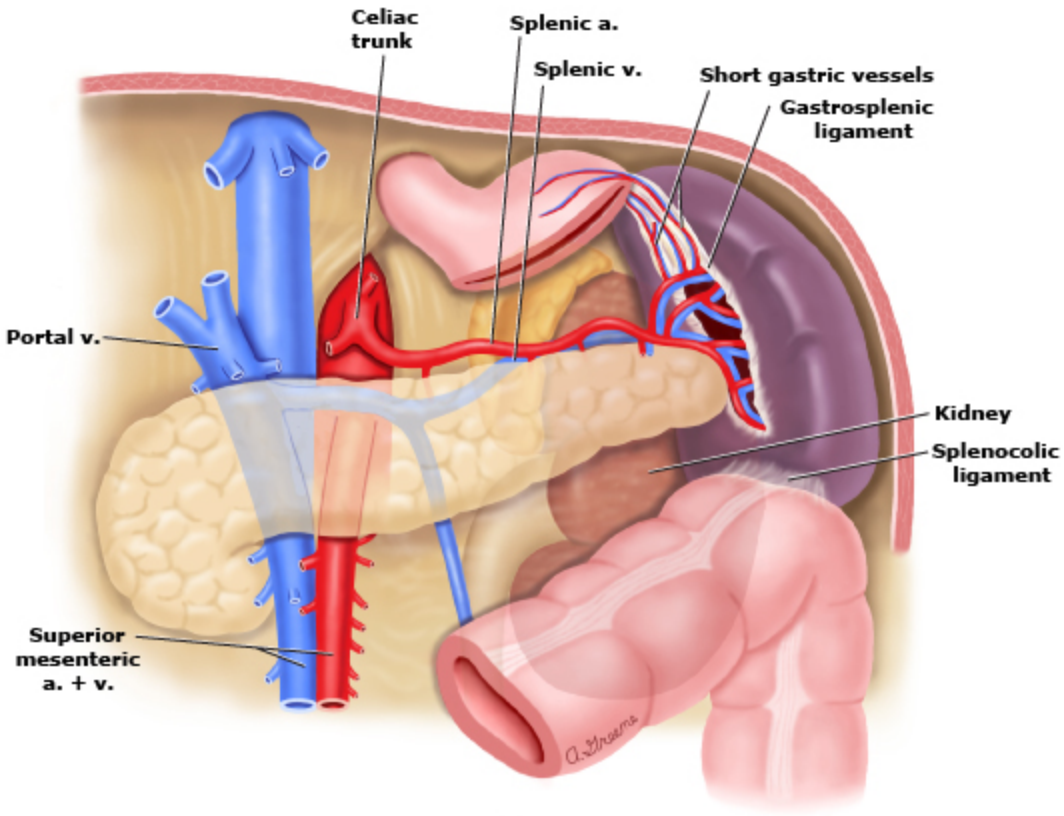
31. Lai L, Brugge WR. Endoscopic ultrasound is a sensitive and specific test to diagnose portal venous system thrombosis (PVST). Am J Gastroenterol 2004; 99:40.
32. Piscaglia F, Gianstefani A, Ravaioli M, et al. Criteria for diagnosing benign portal vein thrombosis in the assessment of patients with cirrhosis and hepatocellular carcinoma for liver transplantation. Liver Transpl 2010; 16:658.
33. Wu M, Schuster M, Tadros M. Update on Management of Portal Vein Thrombosis and the Role of Novel Anticoagulants. J Clin Transl Hepatol 2019; 7:154.
34. Ai MH, Dong WG, Tan XP, et al. Efficacy and safety study of direct-acting oral anticoagulants for the treatment of chronic portal vein thrombosis in patients with liver cirrhosis. Eur J Gastroenterol Hepatol 2020; 32:1395.
35. Nagaoki Y, Aikata H, Daijyo K, et al. Efficacy and safety of edoxaban for treatment of portal vein thrombosis following danaparoid sodium in patients with liver cirrhosis. Hepatol Res 2018; 48:51.
36. Naymagon L, Tremblay D, Zubizarreta N, et al. The Natural History, Treatments, and Outcomes of Portal Vein Thrombosis in Patients With Inflammatory Bowel Disease. Inflamm Bowel Dis 2021; 27:215.
37. Chae HB. The therapeutic effect of davigtran in the liver cirrhosis patients. Hepatol Int 2019.
38. Plessier A. Rivaroxaban Prophylaxis in Noncirrhotic Portal Vein Thrombosis. NEJM Evid 2022; 1.
39. Loffredo L, Pastori D, Farcomeni A, Violi F. Effects of Anticoagulants in Patients With Cirrhosis and Portal Vein Thrombosis: A Systematic Review and Meta-analysis. Gastroenterology 2017; 153:480.
40. Francoz C, Belghiti J, Vilgrain V, et al. Splanchnic vein thrombosis in candidates for liver transplantation: usefulness of screening and anticoagulation. Gut 2005; 54:691.
41. Dumortier J, Vaillant E, Boillot O, et al. Diagnosis and treatment of biliary obstruction caused by portal cavernoma. Endoscopy 2003; 35:446.
42. Simpson WG, Schwartz RW, Strodel WE. Splenic vein thrombosis. South Med J 1990; 83:417.
43. Gouge TH, Ranson JH. Esophageal transection and paraesophagogastric devascularization for bleeding esophageal varices. Am J Surg 1986; 151:47.
44. Borgonovo G, Costantini M, Grange D, et al. Comparison of a modified Sugiura procedure with portal systemic shunt for prevention of recurrent variceal bleeding in cirrhosis. Surgery 1996; 119:214.

45. Luca A, Miraglia R, Caruso S, et al. Short- and long-term effects of the transjugular intrahepatic portosystemic shunt on portal vein thrombosis in patients with cirrhosis. Gut 2011; 60:846.
46. Valentin N, Korrapati P, Constantino J, et al. The role of transjugular intrahepatic portosystemic shunt in the management of portal vein thrombosis: a systematic review and meta-analysis. Eur J Gastroenterol Hepatol 2018; 30:1187.
47. Knight GM, Clark J, Boike JR, et al. TIPS for Adults Without Cirrhosis With Chronic Mesenteric Venous Thrombosis and EHPVO Refractory to Standard-of-Care Therapy. Hepatology 2021; 74:2735.
48. Ganger DR, Klapman JB, McDonald V, et al. Transjugular intrahepatic portosystemic shunt (TIPS) for Budd-Chiari syndrome or portal vein thrombosis: review of indications and problems. Am J Gastroenterol 1999; 94:603.
49. Radosevich PM, Ring EJ, LaBerge JM, et al. Transjugular intrahepatic portosystemic shunts in patients with portal vein occlusion. Radiology 1993; 186:523.
50. Orr DW, Harrisson PM, Karani J, et al. Chronic portomesenteric and portosplenomesenteric venous thrombosis: evaluation of long term follow-up and determinants of survival. Hepatology 2005; 42:212A.

Topic 88014 Version 20.0

GRAPHICS

Vascular supply of the spleen



The splenic artery is a branch of the celiac trunk. It traverses along the superior margin of the pancreas toward the spleen, branching to form up to six major arteries before entering the spleen. The splenic vein joins the superior mesenteric vein to form the portal vein.

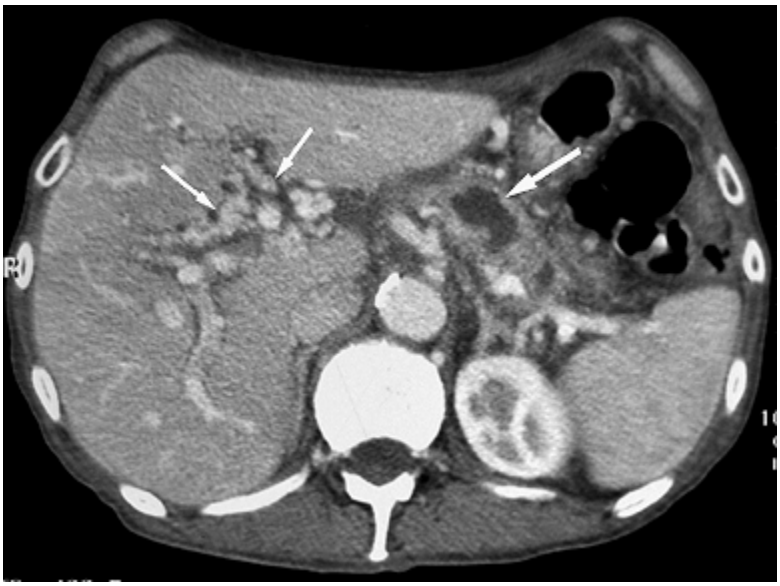
Graphic 68680 Version 4.0

Causes of portal vein thrombosis

Abdominal sepsis
Abdominal surgery
Behçet's syndrome
Cirrhosis
Collagen vascular diseases (eg, lupus)
Compression or invasion of the portal vein by tumor (eg, pancreatic cancer)
Endoscopic sclerotherapy
Hepatocellular carcinoma
Inflammatory bowel disease
Inherited thrombophilias
Myeloproliferative syndromes
Omphalitis
Oral contraceptives
Pancreatic islet cell transplantation
Pancreatitis
Paroxysmal nocturnal hemoglobinuria
Pregnancy
Retroperitoneal fibrosis
Transjugular intrahepatic portosystemic shunt
Trauma

Graphic 73799 Version 5.0

Cavernous transformation of portal vein thrombosis



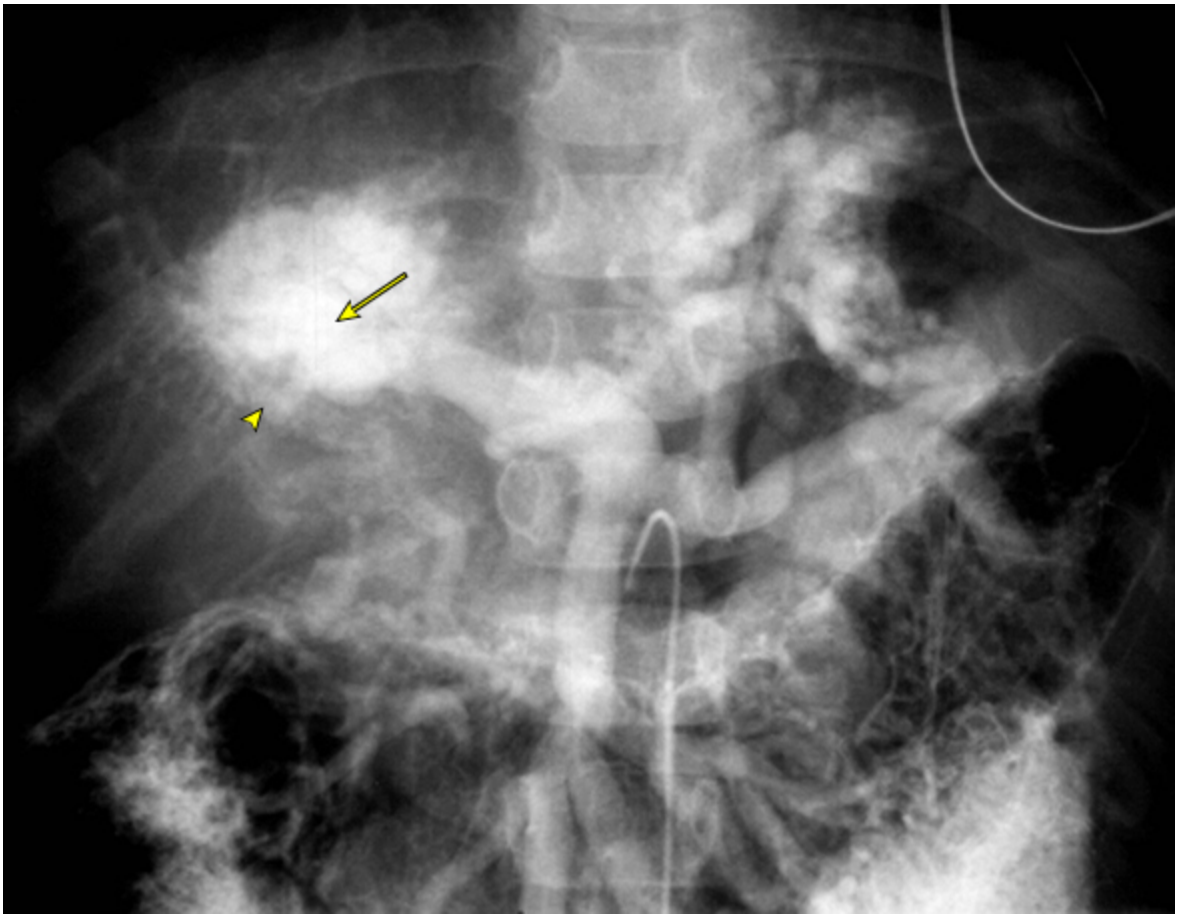
Contrast-enhanced CT scan of the upper abdomen in a 45-year-old male alcoholic demonstrates multiple periportal collateral vessels (small arrows) which have developed secondary to chronic portal vein thrombosis. The portal vein thrombosed due to chronic pancreatitis; a small pancreatic pseudocyst is seen in the body of the pancreas (large arrow).

CT: computed tomography.

Courtesy of Jonathan Kruskal, MD.

Graphic 51167 Version 4.0

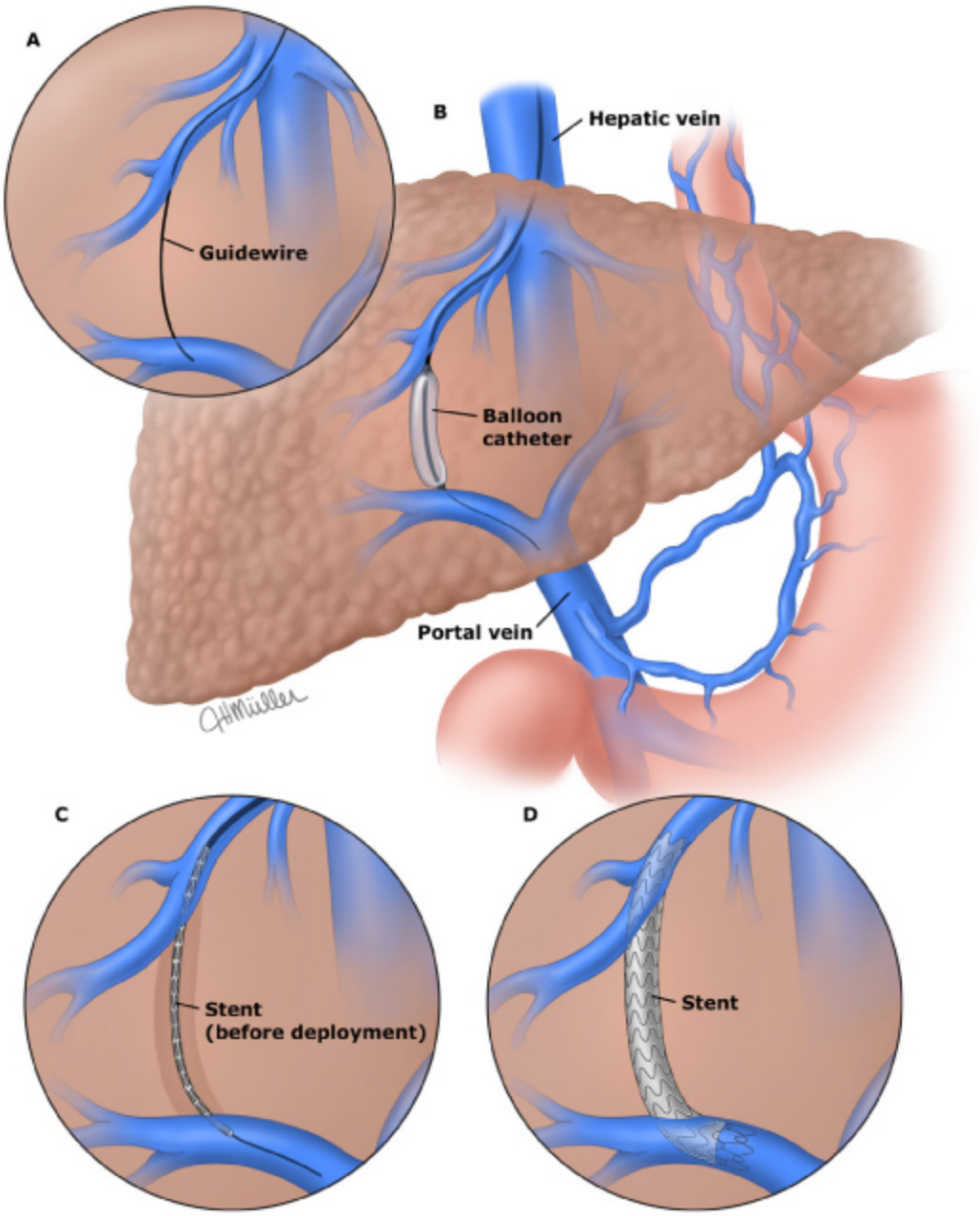
Cavernous transformation of portal vein on arteriography



The venous phase of a mesenteric angiogram shows an occluded portal vein (arrow) with venous collaterals in the porta hepatis (arrowhead).

Graphic 93257 Version 1.0

Transjugular intrahepatic portosystemic shunt



A transjugular intrahepatic portosystemic shunt (TIPS) is created by passing a needle catheter via the transjugular route into the hepatic vein and wedging it there. The needle is then extruded and advanced through the liver parenchyma to the intrahepatic portion of the portal vein and a stent is placed between the portal and hepatic veins. A TIPS functions like side-to-side surgical portacaval shunt, but does not require general anesthesia or major surgery for placement. (A) Passage of a guidewire between the hepatic vein and the portal vein. (B) Inflation of a balloon catheter within the liver to dilate the tract between the hepatic vein and the portal vein. (C) Deployment of the stent. (D) Stent in its final position.

Graphic 72311 Version 2.0

Contributor Disclosures

Arun J Sanyal, MD Equity Ownership/Stock Options: Durect [NASH]; Exhale NZ [Helicobacter pylori]; Genfit [NASH]; HemoShear [Rare liver diseases]; Indalo [NASH]; NorthSea [NASH]; Rivus [NASH]; Sanyal Bio [Animal testing]; Tiziana [NASH]. Grant/Research/Clinical Trial Support: Alnylam [NASH]; Amgen [NASH]; Boehringer Ingelheim [NASH]; Bristol Myers [NASH]; Covance [Lipoproteins]; Echosens Sandhill [NASH]; Fractyl [NASH]; Genentech [NASH]; Gilead [NASH]; HistoIndex [NASH]; Immuron [Alcoholic hepatitis]; Inventiva [NASH]; Lilly [NASH]; Madrigal [NASH]; Mallinckrodt [Portal hypertension]; Merck [NASH]; Novartis [NASH]; Novo Nordisk [NASH]; Owl [NASH]; Path AI [NASH]; Pfizer [NASH]; ProSciento [NASH]; Regeneron [NASH]; Roche [NASH]; Salix [Hepatic encephalopathy]; Second Genome [Microbiome]; Siemens [NASH]. Consultant/Advisory Boards: 89 Bio [NASH]; Albireo [NASH]; Amgen [NASH]; Amra [Metabolism]; AstraZeneca [NASH]; BiocellVia [NASH]; Boehringer Ingelheim [NASH]; Bristol Myers [NASH]; Conatus [NASH]; Fractyl [NASH]; Galectin [NASH]; Genentech [NASH]; Genfit [NASH]; Gilead [NASH, COVID-19]; HemoShear [Rare diseases]; HistoIndex [NASH]; Immuron [Alcohol-associated liver disease]; Intercept [NASH]; Janssen [NASH]; Lilly [NASH]; Madrigal [NASH]; Mallinckrodt [Portal hypertension]; Merck [NASH]; NGM Bio [NASH]; NorthSea [NASH]; Novartis [NASH]; Novo Nordisk [NASH]; PathAI [NASH]; Perspectum [NASH]; Pfizer [NASH]; Poxel [NASH]; ProSciento [NASH]; Regeneron [NASH]; Roche [NASH]; Salix [Cirrhosis]; Sanofi [NASH]; Sequana [Cirrhosis]; Siemens [NASH]; Takeda [NASH]; Terns [NASH]. All of the relevant financial relationships listed have been mitigated. **Sanjiv Chopra, MD, MACP** No relevant financial relationship(s) with ineligible companies to disclose. **Kristen M Robson, MD, MBA, FACG** No relevant financial relationship(s) with ineligible companies to disclose.

Contributor disclosures are reviewed for conflicts of interest by the editorial group. When found, these are addressed by vetting through a multi-level review process, and through requirements for references to be provided to support the content. Appropriately referenced content is required of all authors and must conform to UpToDate standards of evidence.

Conflict of interest policy

→