RadioGraphics

Diagnostic and Interventional Radiology for Budd-Chiari Syndrome[1]

Marco Cura, MD • Ziv Haskal, MD[2] • Jorge Lopera, MD

CME FEATURE

See accompanying test at http://www.rsna.org/education/rg_cme.html

LEARNING OBJECTIVES FOR TEST 1

After reading this article and taking the test, the reader will be able to:

- Describe the pathophysiologic and histopathologic changes of Budd-Chiari syndrome.
- Recognize the indications for a particular method of treatment in patients with this syndrome.
- Explain how a transjugular intrahepatic portosystemic shunt works to treat Budd-Chiari syndrome.

TEACHING POINTS

See last page

Budd-Chiari syndrome is a heterogeneous group of disorders characterized by hepatic venous outflow obstruction that involves one or more draining hepatic veins. Its occurrence in populations in the western hemisphere is commonly associated with hypercoagulative states. Clinical manifestations in many cases are nonspecific, and imaging may be critical for early diagnosis of venous obstruction and accurate assessment of the extent of disease. If Budd-Chiari syndrome is not treated promptly and appropriately, the outcome may be dismal. Comprehensive imaging evaluations, in combination with pathologic analyses and clinical testing, are essential for determining the severity of disease, stratifying risk, selecting the appropriate therapy, and objectively assessing the response. The main goal of treatment is to alleviate hepatic congestion, thereby improving hepatocyte function and allowing resolution of portal hypertension. Various medical, endovascular, and surgical treatment options are available. Percutaneous and endovascular procedures, when performed in properly selected patients, may be more effective than medical treatment methods for preserving liver function and arresting disease progression in the long term. In addition, such procedures are associated with lower morbidity and mortality than are open surgical techniques.

Abbreviations: IVC = inferior vena cava, TIPS = transjugular intrahepatic portosystemic shunt

RadioGraphics 2009; 29:669–681 • **Published online** 10.1148/rg.293085056 • **Content Codes:** GI VI

[1]From the Department of Radiology, University of Texas Health Science Center, 7703 Floyd Curl Dr, Mail Code 7800, San Antonio, TX 78229-3900 (M.C., J.L.); and Department of Radiology, Columbia University Medical Center, New York, NY (Z.H.). Recipient of a Certificate of Merit award for an education exhibit at the 2007 RSNA Annual Meeting. Received March 17, 2008; revision requested April 23 and received August 20; accepted August 21. Z.H. is a member of the advisory boards of W. L. Gore & Associates, AngioDynamics, Elcam, OmniSonics Medical Technologies, and NorthPoint Domain and a consultant for C. R. Bard, Boston Scientific, and NovaShunt, and has received royalties from Microtek International and Cook Group. All other authors have no financial relationships to disclose. **Address correspondence to** M.C. (e-mail: *marcocura@yahoo.com*).

[2]**Current address:** Department of Radiology, University of Maryland, Baltimore, Md.

Introduction

Budd-Chiari syndrome is a heterogeneous group of disorders characterized by hepatic venous outflow obstruction at the level of the hepatic veins, the inferior vena cava (IVC), or the right atrium (1). Budd-Chiari syndrome is not a primary condition of the liver parenchyma; it is the result of partial or complete obstruction of hepatic venous outflow.

Hepatic venous outflow obstruction increases hepatic sinusoidal pressure. The increased pressure results in portal hypertension, liver congestion, and decreased liver perfusion. Liver congestion causes hepatocyte dysfunction, which may progress to hepatocyte necrosis and hepatic fibrosis and cirrhosis. Budd-Chiari syndrome is a pathologic disorder with multiple causes. In the western hemisphere, most patients in whom this syndrome develops have an underlying condition that predisposes them to thrombosis. Membranous weblike obstruction of the hepatic vein or IVC, and malignant tumors that obstruct hepatic venous outflow, may lead to Budd-Chiari syndrome.

The manifestations range from mild symptoms to fulminant acute liver failure to chronic disease (2,3). The patient's symptoms and signs at clinical presentation depend on the extension and acuteness of hepatic venous obstruction and the adequacy of the remaining primary and collateral venous outflow (4,5).

Clinical and imaging evaluations and histologic analyses of liver biopsy specimens obtained in patients in whom the presence of Budd-Chiari syndrome is suspected play an important role in the detection and classification of the disease, helping guide case management at presentation and providing valuable information at post-treatment follow-up.

Medical treatment of Budd-Chiari syndrome, which includes anticoagulant therapy to prevent recurrence and extension of thrombosis, along with other measures to control ascites and gastrointestinal bleeding, may alleviate patients' symptoms temporarily but is associated with poor long-term outcomes (6–8). Surgical treatments of Budd-Chiari syndrome involve either the creation of a portosystemic shunt or liver transplantation. Transplantation is reserved for patients with advanced hepatocellular dysfunction, and its feasibility depends on the availability of a donor. Surgical creation of portosystemic shunts, with the exception of the mesoatrial shunt, provides partial decompression of the portal venous system (7,9,10). By contrast, percutaneous interventions can help restore hepatic venous outflow in selected patients, leading to resolution of liver congestion, improving liver function, and arresting hepatic destruction, with a lower mortality than that incurred by surgical procedures.

Etiology and Clinical Manifestations

In approximately 75% of patients, a hematologic abnormality or a cause of thrombotic diathesis can be identified that predisposes the patient to the occurrence of Budd-Chiari syndrome. The presence of multiple causes in the same patient has been reported (11). Hematologic diseases, especially myeloproliferative disorders, are the most common cause, and it has been suggested that patients with idiopathic Budd-Chiari syndrome may have an underlying myeloproliferative disorder (12). Causes of thrombotic diathesis that have been associated with Budd-Chiari syndrome include paroxysmal nocturnal hemoglobinuria, antiphospholipid syndrome, inherited deficiencies of proteins C and S and antithrombin III, factor V Leiden mutation, prothrombin gene mutation, methylene tetrahydrofolate reductase mutation, use of oral contraceptives, pregnancy, and immediate postpartum status (2,6,11,13–24).

Metastatic invasion of the hepatic vein, IVC, or right atrium and primary tumor occurrence in the kidney, liver, adrenal gland, IVC, or heart are less frequent causes of Budd-Chiari syndrome (25–28). Membranous weblike obstruction of the hepatic vein or IVC is a more prevalent cause of hepatic venous outflow obstruction in the Asian population (29). Membranous webs of the hepatic vein or IVC may be congenital or represent sequelae of thrombosis (30,31).

Budd-Chiari syndrome commonly occurs in women and young adults (32,33). The clinical manifestations of Budd-Chiari syndrome depend on both the extension and the acuteness of the venous outflow obstruction. The severity of symptoms and liver dysfunction depends on the efficacy of the remaining primary hepatic venous outflow and the development of venous collateral circulation (4,5). The clinical manifestations at patient presentation range from mild symptoms to signs of fulminant liver failure (2,3). The duration of symptoms, severity of manifestations, and rate of disease progression have been used to differentiate among fulminant, acute, subacute, and chronic forms of Budd-Chiari syndrome (5,9,19,34,35). Some patients with a chronic form may experience an acute exacerbation (36). The subacute form, which manifests with portal hypertension, ascites, and liver failure with varying degrees of severity, is the most common.

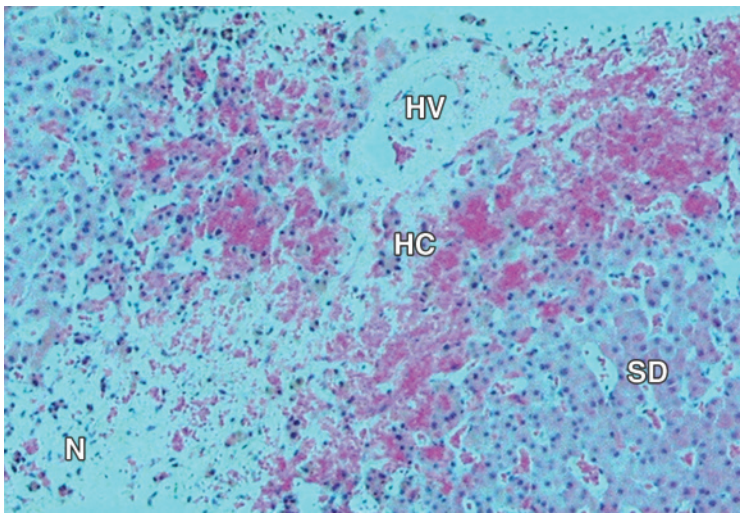


Figure 1. Photomicrograph (original magnification, ×25; hematoxylin-eosin stain) shows a liver specimen obtained at transjugular biopsy before transjugular intrahepatic portosystemic shunt (TIPS) creation in a 43-year-old woman with Budd-Chiari syndrome. The specimen contains areas of hepatic congestion *(HC)*, necrosis *(N)* characterized by poorly stained and mummified hepatocytes with lysed nuclei, sinusoidal dilatation *(SD)*, and clustering of red blood cells in the sinusoidal spaces around the hepatic vein *(HV)*, in the perivenular zone.

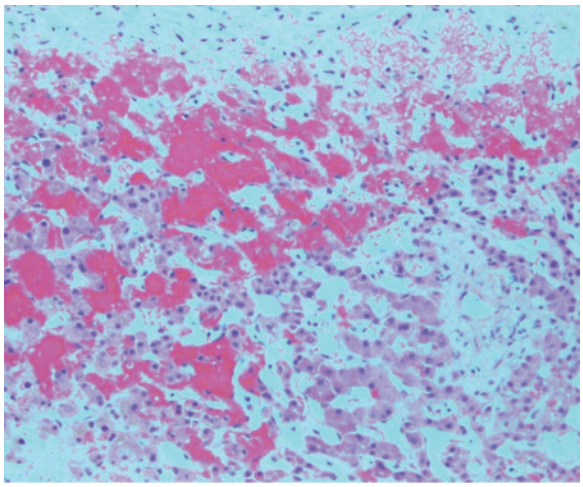


Figure 2. Photomicrograph (original magnification, ×40; hematoxylin-eosin stain) of a transjugular liver biopsy specimen from a 27-year-old woman with acute Budd-Chiari syndrome shows sinusoidal dilatation and clustering of red blood cells in the sinusoidal spaces around the hepatic vein, findings indicative of congestion in the perivenular zone.

Jaundice and encephalopathy are more common in fulminant and acute forms of the syndrome, and splenomegaly and esophageal and gastric varices are predominant in the chronic form.

Pathophysiology

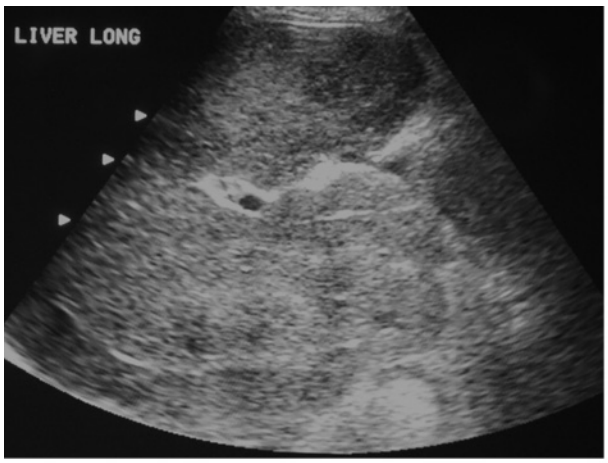
Teaching Point

Inability of blood flow to drain from the liver leads to increased sinusoidal pressure and hepatic congestion. The obstruction of hepatopetal portal venous flow causes portal hypertension. The ensuing venous stasis increases the sinusoidal pressure, leading to a reduction of portal perfusion (6). The decreased hepatic perfusion and resultant congestion cause ischemic injury to liver cells that culminates in the development of hepatocyte necrosis in the perivenular zones (37,38). Hepatocyte necrosis is followed by progressive fibrosis, nodular regenerative hyperplasia, and ultimately liver cirrhosis.

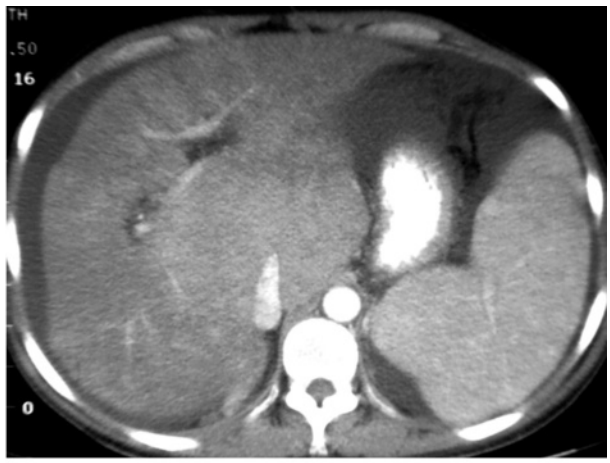
In chronic cases, the development of bridging intrahepatic and capsular venovenous collateral vessels may permit the egress of venous flow (39). If sinusoidal pressure is not reduced and hepatic congestion is not relieved by the development of portal venous collaterals or the creation of portosystemic shunts, Budd-Chiari syndrome progresses to eventual liver failure (6,32).

Pathologic Features

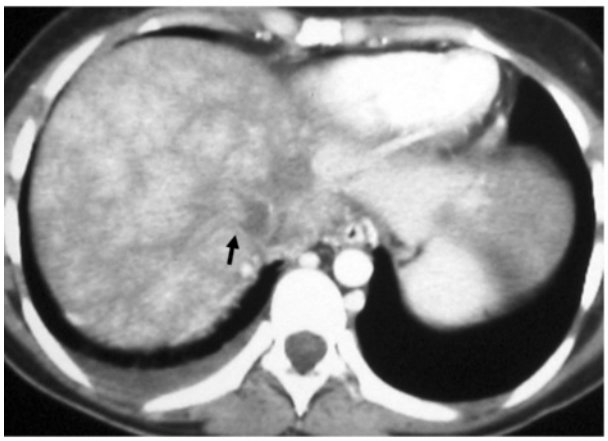
In the early phase of disease, hepatic venous congestion begins around the hepatic veins, in the centrilobular or perivenular zone (4,23). The cord–sinusoid architecture is preserved, but there is sinusoidal dilatation, and red blood cells are clustered in the sinusoids and the Disse space (perisinusoidal zone) (40,41). The process then extends to the middle zone and, from there, to the periportal zone of the hepatic lobule (23) (Figs 1, 2). In severely congested areas, there is a decrease in liver perfusion, which results in anoxic damage and necrosis of hepatocytes. Necrosis occurs with a zonal distribution pattern, starting in the perivenular zone. Depending on whether only hepatic veins or both hepatic and portal veins are obstructed, necrosis of contiguous liver cells may span adjacent lobules in a central-to-central or central-to-portal fashion. Later, necrotic hepatocytes are replaced by fibrosis, which begins with the proliferation of Ito cells in the perivenular and perisinusoidal zones. Bridging fibrosis is seen in a venovenous "reversed-lobulation" pattern or a portovenous or portoportal distribution (42).



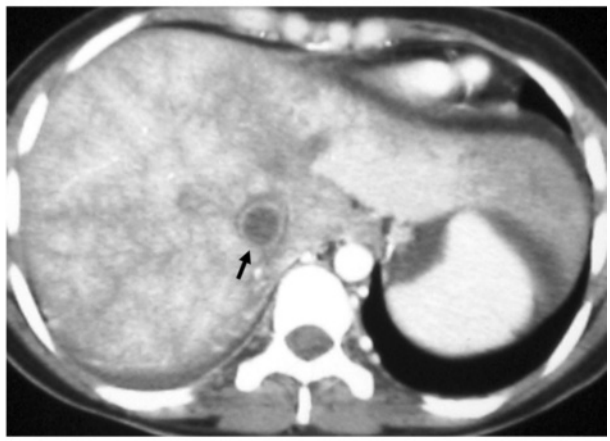
3a.



3b.



4a.



4b.

Figures 3, 4. US and CT features of Budd-Chiari syndrome. **(3a)** Gray-scale US image obtained in a 55-year-old woman shows a globally enlarged liver and heterogeneity of the hepatic parenchyma. **(3b)** Contrast material–enhanced abdominal CT image shows ascites and stronger enhancement in the caudate lobe and central portion of the liver parenchyma than in the periphery. **(4a, 4b)** Contrast-enhanced abdominal CT images obtained in a 30-year-old woman (**4a** at a higher level than **4b**) show patchy enhancement of the liver parenchyma, hypertrophy of the left hepatic lobe, and thrombosis of the hepatic veins and IVC (arrow).

Multiple obstructions of hepatic venous outflow often occur asynchronously, causing various degrees of liver injury in different hepatic regions (43).

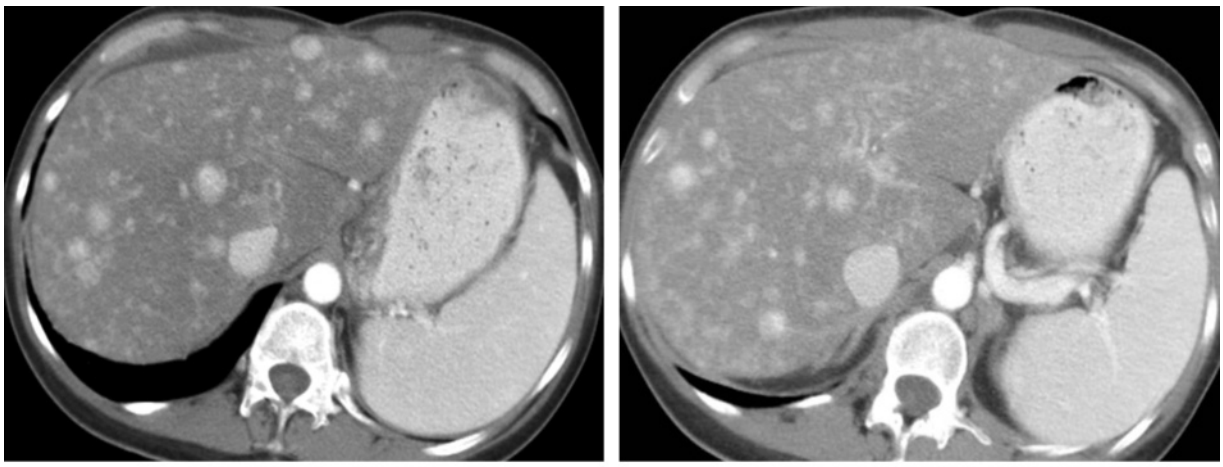
In patients with long-standing Budd-Chiari syndrome, florid cirrhosis occurs with nodular regeneration and reduced hepatic parenchymal volume. Large regenerative nodules may develop in response to focal losses of portal perfusion and compensatory increases in hepatic arterial flow (44,45).

Imaging Characteristics

Budd-Chiari syndrome has variable imaging features. Hepatic vein or IVC thrombosis, with resultant changes in liver morphology and enhancement patterns, venous collaterals, varices, and ascites may be directly observed. Imaging also plays an important role in the detection and classification of disease by enabling the correlation of anatomic changes with the duration of the pathologic process.

Duplex Doppler ultrasonography (US) is a useful method for detecting Budd-Chiari syndrome because it allows easy assessment of hepatic venous flow and detection of hepatic parenchymal heterogeneity (46–48). Computed tomography (CT) and magnetic resonance (MR) imaging also can depict hepatic venous flow or thrombosis and IVC compression or occlusion (Figs 3, 4).

In the presence of acute disease, the imaging features correspond with histologic findings of liver congestion and edema. The liver is globally enlarged, with lower attenuation on CT images, decreased signal intensity on T1-weighted MR images, and heterogeneously increased signal intensity on T2-weighted MR images, predominantly in the periphery (49). Differential contrast



a. b.

Figure 5. Contrast-enhanced helical CT images from late hepatic arterial phase imaging in a 47-year-old man with Budd-Chiari syndrome show multiple regenerative nodules with marked homogeneous enhancement and, in **a**, ascites.

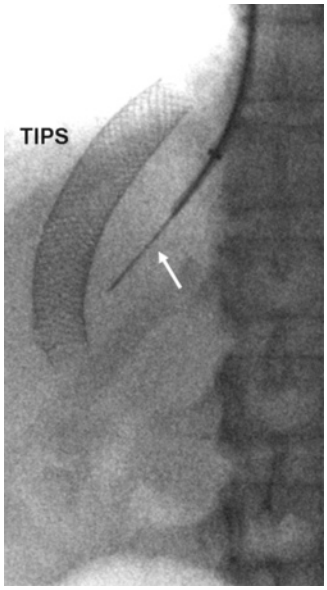


Figure 6. Transcaval transjugular liver biopsy in a 29-year-old woman with Budd-Chiari syndrome after TIPS creation. Spot image shows an automated core needle (arrow) before the outer cutting cannula was fired over the slotted stylet to collect a tissue sample. Three passes were made with the needle to ensure collection of an adequate amount of hepatic tissue and a representative specimen.

enhancement between central and peripheral areas of liver parenchyma is a feature of acute Budd-Chiari syndrome (Fig 4). The more edematous and congested peripheral regions demonstrate decreased contrast enhancement, whereas stronger enhancement is seen in the central parenchyma. After the administration of contrast material, increased enhancement is seen in areas of venous drainage that are less affected, such as the caudate lobe (39,50,51).

The development of intra- and extrahepatic collateral veins in subacute Budd-Chiari syndrome permits the egress of venous flow, producing a more homogeneous enhancement pattern with persistent signs of edema (52,53).

In chronic Budd-Chiari syndrome, there is atrophy of the affected portions of the liver, and the parenchymal edema is replaced by fibrosis, which results in decreased T1- and T2-weighted signal intensity at unenhanced MR imaging and in delayed enhancement in contrast-enhanced studies (39,49,54). Intrahepatic and subcapsular collateral veins are well developed and easily appreciated in the chronic disease phase. Hypertrophy of the caudate lobe, irregularities of the liver contour, and regenerative nodules are prominent features of chronic Budd-Chiari syndrome (51,55,56) (Fig 5).

Liver Biopsies

Teaching Point

Consecutive liver biopsies in patients with Budd-Chiari syndrome are useful for assessing the severity of disease (congestion, necrosis, fibrosis, and regenerative nodules) and determining whether it has progressed after venous decompression procedures. Because of the inhomogeneous distribution of disease in the liver, a single biopsy result is less significant than the trend of histopathologic changes seen in consecutive liver biopsy specimens (57). Specimens obtained after adequate treatment (treatment that restores sufficient liver venous outflow) may exhibit improvement in, or total resolution of, liver congestion.

Many patients with Budd-Chiari syndrome are undergoing anticoagulant therapy or have perihepatic ascites, conditions that are relative contraindications for percutaneous liver biopsy. Transjugular liver biopsy is safer in this group of patients. Venographic and hemodynamic evaluations of affected areas of the liver may be carried out at the same time. A transcaval approach may be used in transjugular liver biopsy when the hepatic vein is not patent (Fig 6).

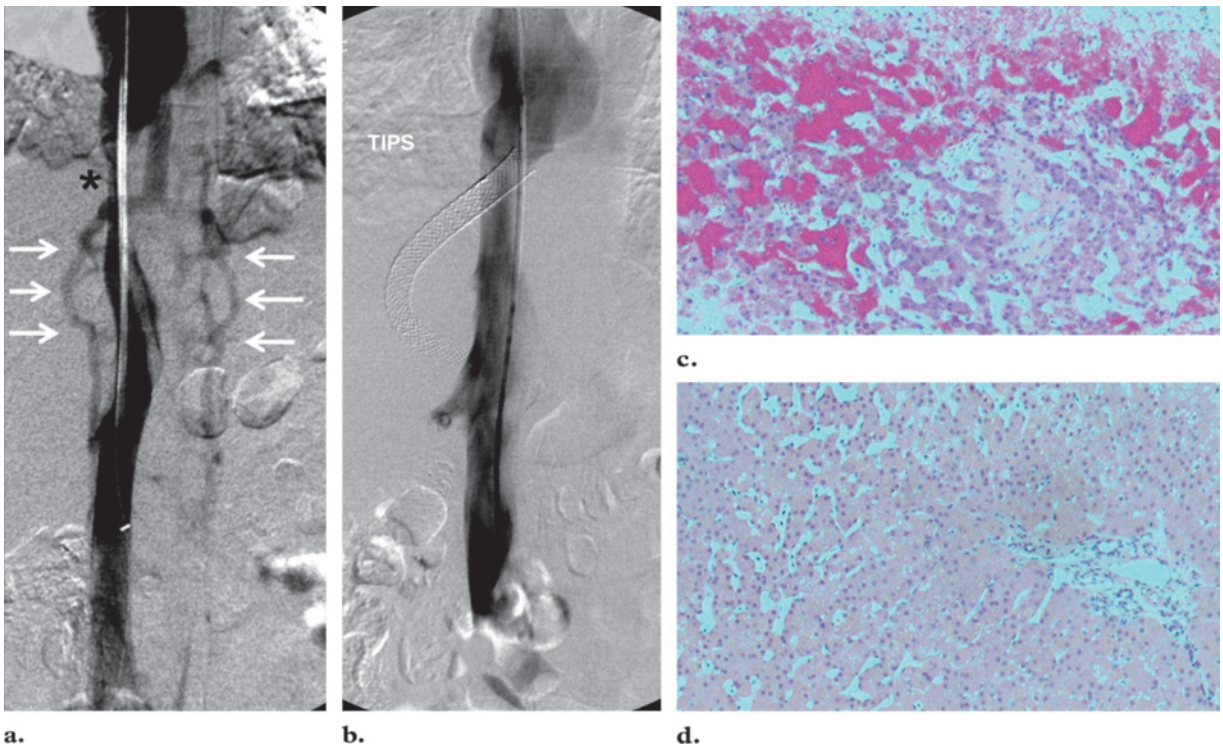


Figure 7. **(a)** Cavogram obtained before TIPS creation in a 24-year-old woman with Budd-Chiari syndrome and a history of oral contraceptive use depicts a reversible intrahepatic IVC stenosis (*). The presence of perilumbar and azygos collateral veins (arrows) is indicative of hemodynamically significant caval narrowing. **(b)** Cavogram obtained 2 months after TIPS creation shows resolution of the intrahepatic IVC stenosis and decompression of venous outflow, findings that correlated with a decrease in the portosystemic gradient from 20 to 6 mm Hg. **(c, d)** Comparison of photomicrographs (original magnification, ×20; hematoxylin-eosin stain) of liver biopsy specimens obtained before **(c)** and 2 months after **(d)** TIPS creation shows posttreatment resolution of perivenular congestion with residual sinusoidal dilatation.

Tests of Metabolic Function

Serum markers of liver function correlate with hepatocyte function. Liver function tests are useful to evaluate disease progression and the effect of therapy. An improvement in liver congestion correlates with a recovery in hepatocyte function. Normalization of albumin and bilirubin values indicates an improvement in liver synthetic and excretory functions.

IVC Involvement

Teaching Point

IVC stenosis and occlusion secondary to liver congestion and compensatory caudate lobe hypertrophy are common in Budd-Chiari syndrome. Cavograms in patients with the syndrome commonly demonstrate stenosis of the intrahepatic segment of the IVC. The presence of collateral cavocaval circulation is an indication of the effect of stenosis on caval hemodynamics (23). IVC stenosis is reversible and may resolve when hepatic congestion has been relieved by successful venous decompression (Fig 7).

Disease Outcomes

Unless hepatic congestion is relieved, the disease process in Budd-Chiari syndrome eventually leads to liver failure and death (32). Progressive hepatic decompensation and fibrosis are expected in patients with liver congestion (35). However, a working portosystemic shunt corrects hemodynamic abnormalities, allowing venous decompression, relief of liver congestion, and reversal of liver damage, with resultant improvements in liver function, quality of life, and survival (23,35) (Fig 7c, 7d).

Treatment Options

Teaching Point

The primary goal of treatment is the resolution of hepatic congestion in order to improve liver perfusion and preserve functioning hepatocytes.

The available therapies for Budd-Chiari syndrome are diverse, and factors that influence the prognosis should be considered when stratifying risks and selecting a therapy (36,58). The selection of a specific treatment method is directed by whether the predominant manifestation is liver failure or portal hypertension. It also depends on the potential for parenchymal recovery, the surgical risk, and the availability of a liver donor (43).

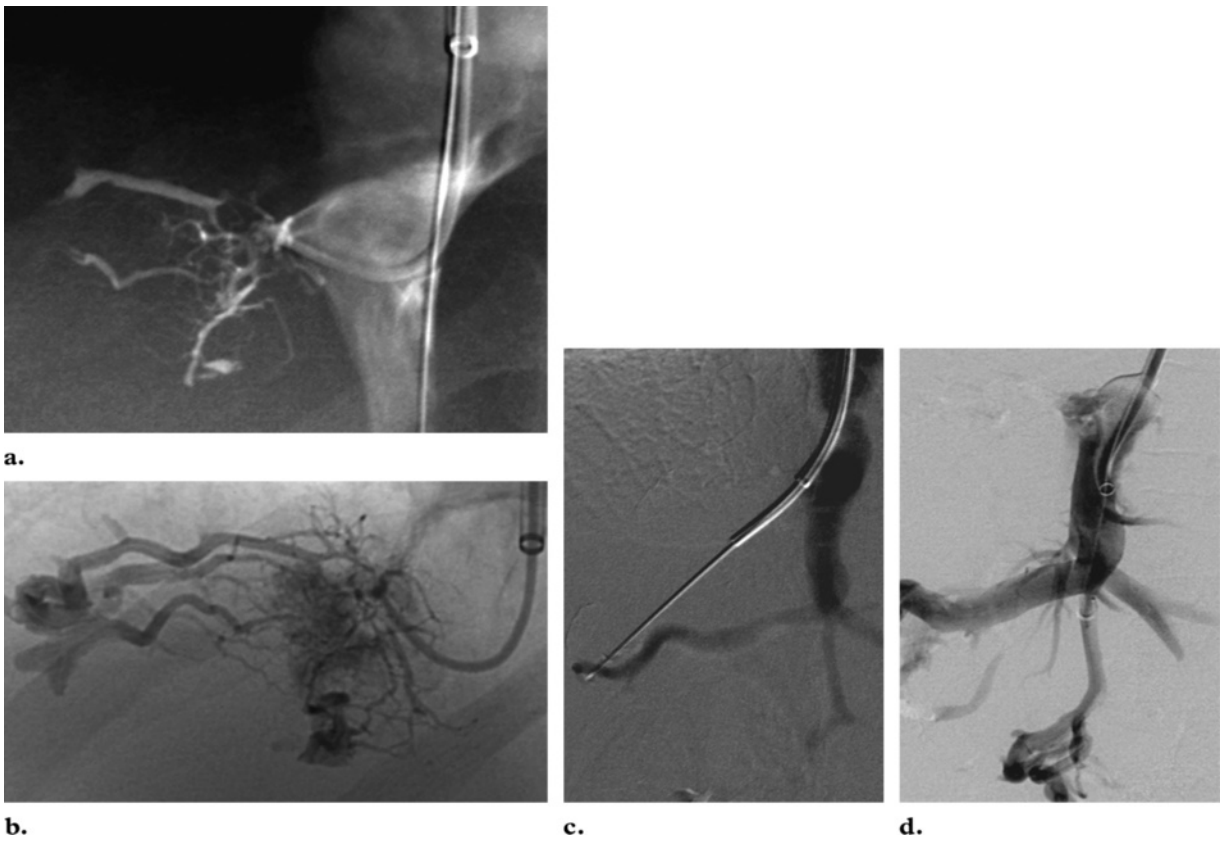


Figure 8. **(a, b)** Initial venograms obtained in a 33-year-old woman with Budd-Chiari syndrome due to a factor V Leiden genetic mutation and with abdominal pain show occlusion of the right hepatic vein and spiderweblike intrahepatic venous collaterals. **(c)** Transcaval CO_2 wedge venogram shows an intrahepatic collateral vein draining into the middle hepatic vein. **(d)** Venogram obtained with selective catheterization of the middle hepatic vein shows ostial stenosis and collateral veins.

Medical management of Budd-Chiari syndrome may be offered to patients with mild symptoms and no evidence of liver necrosis (12). It consists of control of ascites, prevention of further venous thrombosis, and management of any underlying condition. Ascites is treated with a low sodium intake, spironolactone, and furosemide. Large-volume paracentesis with albumin infusion is reserved for cases of refractory ascites (59). Anticoagulant therapy is accomplished with the administration of intravenous heparin initially and then with oral warfarin or subcutaneous low-molecular-weight heparin. Medical treatment has been considered ineffective in most patients and is associated with poor long-term results (6–8). It may control patients' symptoms temporarily, but it fails to arrest the progression of disease.

The development of other treatment options, such as portosystemic shunt creation and liver transplantation, has expanded the therapeutic algorithm. Liver transplantation may reverse certain underlying inborn errors of metabolism (eg, antithrombin III deficiency) (60) and may be the treatment of choice in cases of fulminant liver failure or advanced liver cirrhosis. However, because of the limited number of liver donors and the need for timely intervention, shunt creation is more often performed.

Recanalization of stenotic or occluded hepatic veins to restore venous outflow is the initial procedure of choice. Patent hepatic vein segments with central and focal stenosis or occlusion are best treated with angioplasty, stent placement, or both (61). Various approaches may be used to access the hepatic veins. Transjugular catheterization is attempted first. If the stenotic or occluded hepatic vein cannot be crossed by using a transjugular approach, a US-guided transhepatic needle puncture of a patent segment of the hepatic vein is performed to obtain access. A wire is inserted at the transhepatic access site, used to penetrate the venous obstruction, and then snared and pulled out through the jugular venous access (62). Angioplasty and stent placement then may be performed via the jugular vein to avoid enlarging the site of liver capsule puncture (Figs 8–10).

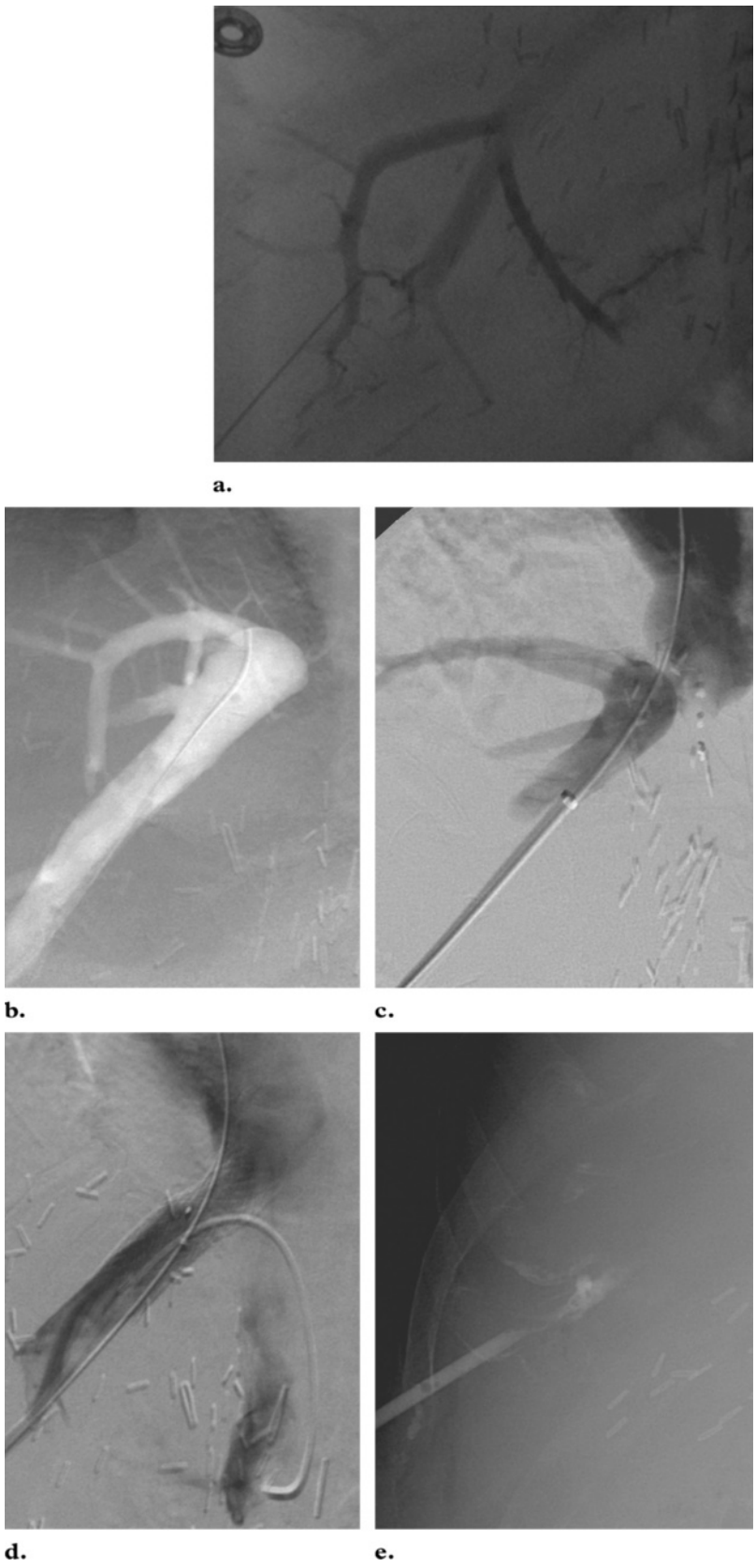
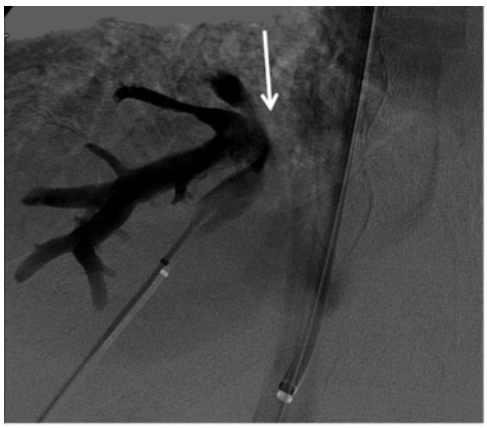
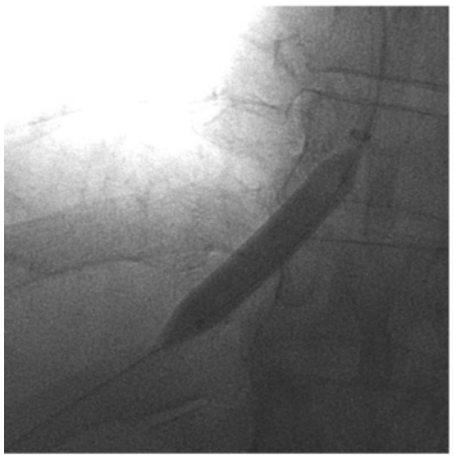


Figure 9. Hepatic vein stenosis in a 35-year-old woman with abdominal pain and ascites due to Budd-Chiari syndrome after liver transplantation. Because of its severity, the stenosis could not be crossed via the jugular vein and had to be accessed percutaneously with a transhepatic approach. Hepatic venograms show the insertion of a 21-gauge needle into the hepatic vein **(a)**, the absence of contrast material reflux into the IVC **(b)**, subsequent retrograde access toward the IVC **(c)**, deployment of a balloon-expandable stent **(d)**, and embolization of the hepatic needle tract with a gelatin sponge slurry **(e)**.

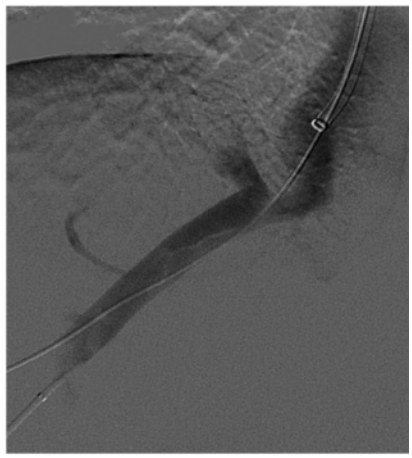
Local pharmacologic thrombolysis, in combination with mechanical thrombolysis, is useful for treating acute or subacute thrombosis of the hepatic vein and IVC. A local infusion of thrombolytic agents is clearly beneficial if new thrombi form after interventional procedures have restored an appreciable amount of flow



a.

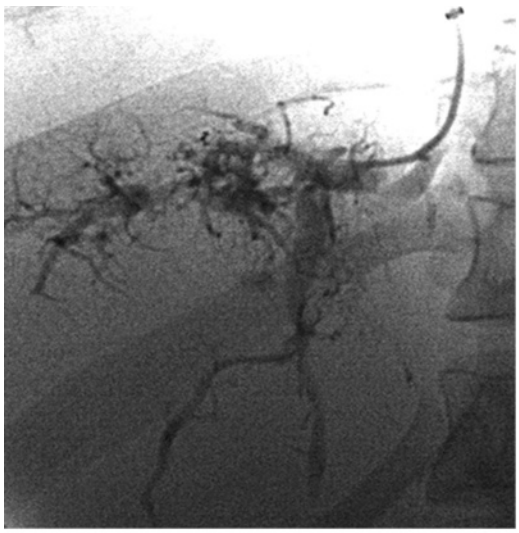


b.

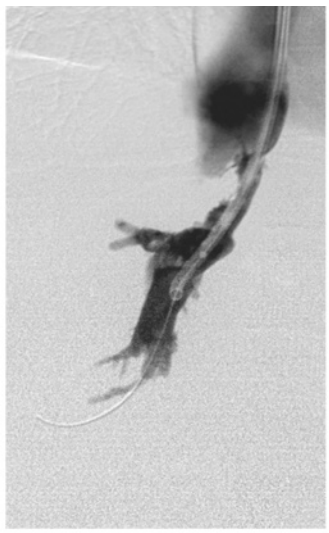


c.

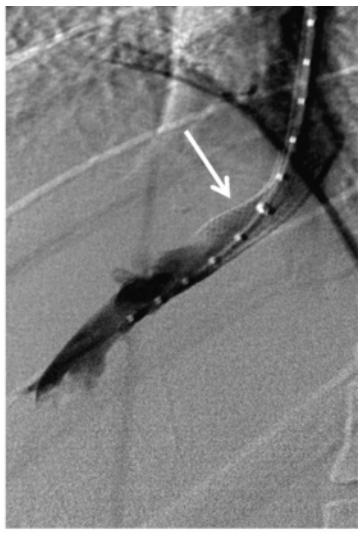
Figure 10. Hepatic vein occlusion in a 42-year-old man with ascites and abdominal pain due to Budd-Chiari syndrome. **(a)** Hepatic venogram shows the occluded vein (arrow), which could not be accessed via the jugular vein and had to be accessed percutaneously with a transhepatic approach. **(b, c)** Comparison of hepatic venograms obtained before **(b)** and after **(c)** angioplasty shows a posttreatment increase in the luminal diameter of the vein.



a.



b.



c.

Figure 11. Hepatic venograms obtained in a 33-year-old woman with Budd-Chiari syndrome after recent angioplasty of the hepatic vein. In **a**, subacute thrombosis of the hepatic vein is depicted. After pharmacomechanical thrombolysis with a pulse-spray infusion of tissue plasminogen activator **(b)**, repeat angioplasty of the ostial stenosis was performed and a stent (arrow in **c**) was placed in the vein.

through the occluded hepatic veins (63) (Fig 11). Focal stenoses of the hepatic vein and web-like occlusions of the hepatic vein and the IVC respond well to angioplasty. Stents are reserved for use in cases in which there is significant residual or recurrent stenosis or occlusion after angioplasty.

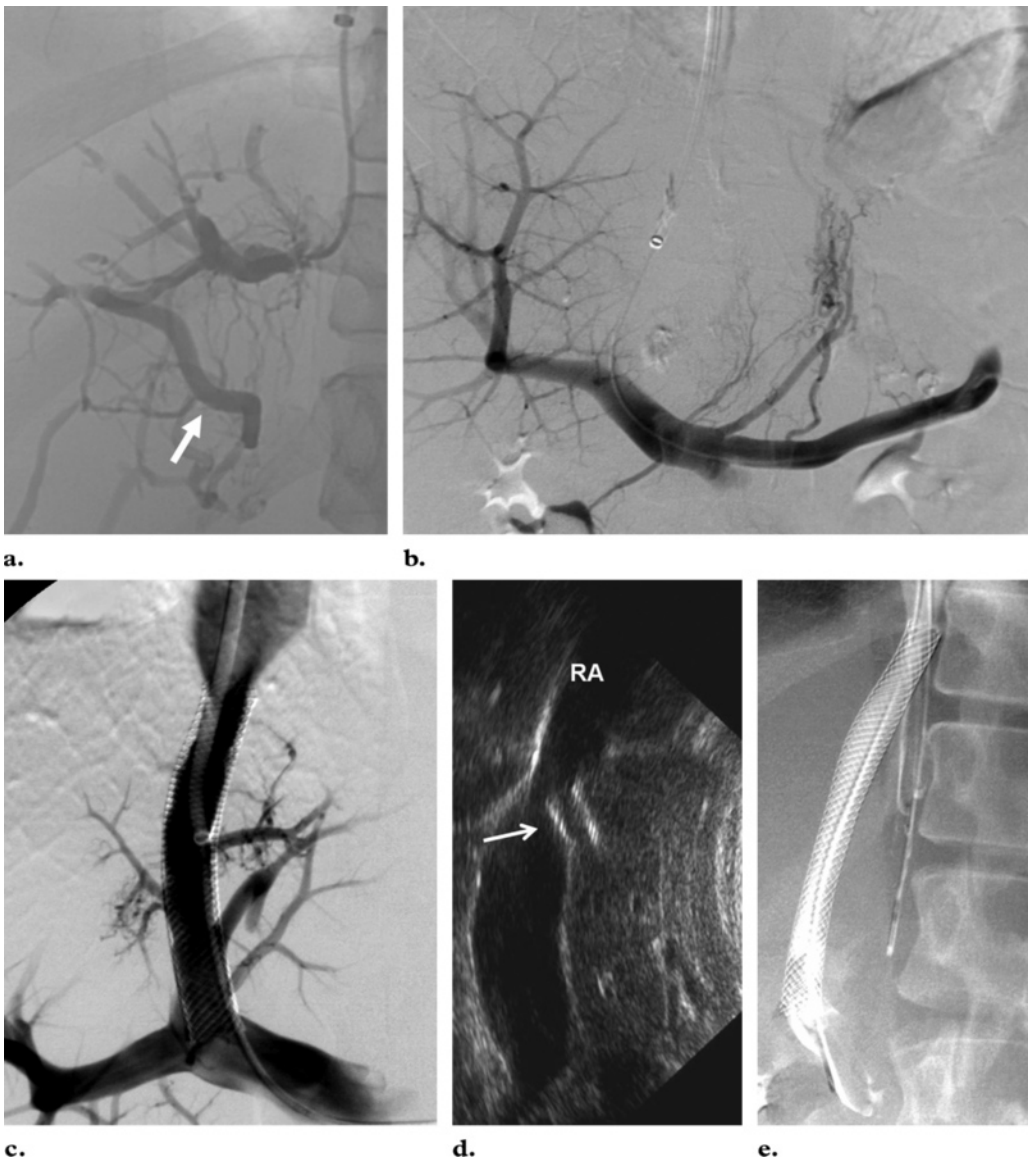
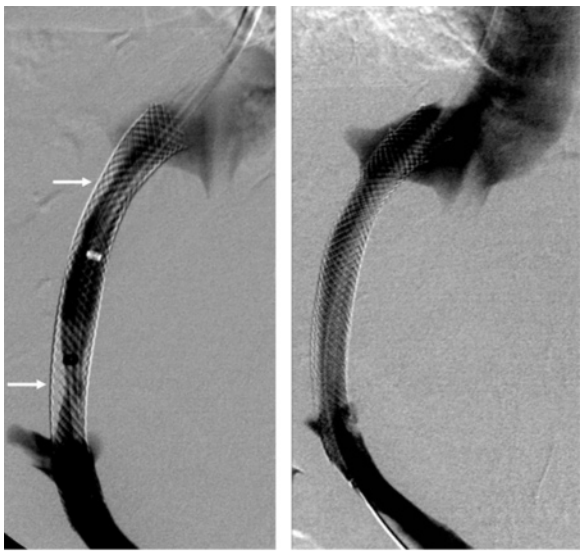


Figure 12. TIPS creation for treatment of Budd-Chiari syndrome in a 53-year-old woman with a history of myelodysplastic syndrome. **(a)** Venogram shows an occluded right hepatic vein (arrow) and multiple intrahepatic collateral veins. **(b, c)** Portal venograms depict access to the portal vein **(b)** for creation of a TIPS **(c)**. **(d, e)** US image **(d)** and venogram **(e)** show hepatic venous outflow via the TIPS into the IVC (arrow) and then without obstruction to the right atrium *(RA)*.

Caval stenosis secondary to liver congestion resolves after successful venous decompression procedures (Fig 7). By contrast, a primary IVC stenosis or occlusion that affects hepatic venous outflow must be addressed to improve the hepatic venous outflow. Angioplasty of the IVC may be performed with a jugular or femoral approach. Percutaneous mechanical and pharmacologic methods of thrombolysis are effective for treating an associated caval thrombosis. Angioplasty of the IVC stenosis is performed after successful thrombolysis, to minimize the risk of pulmonary embolization (64). If angioplasty fails, a caval stent is placed.



a. b.

Figure 13. Recurrent stenosis in a 56-year-old woman with ascites after TIPS creation for treatment of Budd-Chiari syndrome. **(a)** Venogram depicts stenoses (arrows) of the TIPS lumen. **(b)** Venogram obtained after angioplasty demonstrates improved luminal diameter and flow.

Teaching Point

In patients in whom the hepatic vein cannot be recanalized or in whom recanalization produces inadequate liver decongestion, TIPS creation is a good alternative method for relieving liver congestion, with lower mortality and morbidity than those associated with open surgical procedures (65,66). TIPS creation in patients with Budd-Chiari syndrome was initially described by Ochs et al (66) (Fig 12). Not only is the procedure less invasive than open surgery, but a TIPS also has the advantage over side-to-side portocaval, mesocaval, and splenorenal portosystemic shunts because it bypasses the caval stenosis. Like a mesoatrial shunt, a TIPS provides venous decompression by allowing outflow into the suprahepatic IVC, at a site proximal to the intrahepatic stenosis.

Successful TIPS creation is followed by progressive improvements in clinical symptoms, liver function, and liver congestion and by arrested progression of liver fibrosis (6,10,65). Improvement can be seen even in patients with chronic Budd-Chiari syndrome (6,8,10,65–67). On the other hand, surgically created portocaval shunts, with the exception of mesoatrial shunts, may not provide sufficient decompression of the portal venous system, because they drain only the region proximal to the IVC stenosis. Moreover, surgical shunt creation has been associated with higher mortality (7,9,10). The presence of a surgical shunt, especially one in which access to the splanchnic circulation has been accomplished by means of a direct connection with the portal vein, complicates liver transplantation and may increase blood loss and mortality (32,68).

The advantage of a TIPS over most surgically created portosystemic shunts in patients with Budd-Chiari syndrome is that the TIPS allows venous decompression irrespective of the presence of an IVC obstruction.

TIPS Revision

In patients with Budd-Chiari syndrome, TIPS dysfunction used to be fairly common (69–71). These patients' hypercoagulability predisposes them to shunt thrombosis and occlusion. However, an overall improvement in TIPS patency with the use of expanded polytetrafluoroethylene (ePTFE)-coated stents has benefited patients with Budd-Chiari syndrome (64,72) (Fig 13). Better TIPS patency rates have been achieved in this group of hypercoagulable patients by using ePTFE-covered stents than by using bare stents (73).

Summary

Budd-Chiari syndrome is induced by hepatic venous outflow obstruction, which leads to progressive sinusoidal congestion, centrilobular necrosis, fibrosis, and nodular regeneration. Percutaneous therapies performed in properly selected patients can help improve liver function and arrest hepatic destruction.

References

1. Ludwig J, Hashimoto E, McGill DB, van Heerden JA. Classification of hepatic venous outflow obstruction: ambiguous terminology of the Budd-Chiari syndrome. Mayo Clin Proc 1990;65(1):51–55.
2. Hadengue A, Poliquin M, Vilgrain V, et al. The changing scene of hepatic vein thrombosis: recognition of asymptomatic cases. Gastroenterology 1994; 106(4):1042–1047.
3. Valla DC. Hepatic vein thrombosis (Budd-Chiari syndrome). Semin Liver Dis 2002;22(1):5–14.
4. Parker RG. Occlusion of the hepatic veins in man. Medicine (Baltimore) 1959;38:369–402.
5. Dilawari JB, Bambery P, Chawla Y, et al. Hepatic outflow obstruction (Budd-Chiari syndrome): experience with 177 patients and a review of the literature. Medicine (Baltimore) 1994;73(1):21–36.

6. Perelló A, García-Pagán JC, Gilabert R, et al. TIPS is a useful long-term derivative therapy for patients with Budd-Chiari syndrome uncontrolled by medical therapy. Hepatology 2002;35(1):132–139.
7. Kugelmas M. Budd-Chiari syndrome: treatment options and the value of liver transplantation. Hepatogastroenterology 1998;45(23):1381–1386.
8. Blum U, Rössle M, Haag K, et al. Budd-Chiari syndrome: technical, hemodynamic, and clinical results of treatment with transjugular intrahepatic portosystemic shunt. Radiology 1995;197(3):805–811.
9. Bismuth H, Sherlock DJ. Portasystemic shunting versus liver transplantation for the Budd-Chiari syndrome. Ann Surg 1991;214(5):581–589.
10. Mancuso A, Fung K, Mela M, et al. TIPS for acute and chronic Budd-Chiari syndrome: a single-centre experience. J Hepatol 2003;38(6):751–754.
11. Denninger MH, Chaït Y, Casadevall N, et al. Cause of portal or hepatic venous thrombosis in adults: the role of multiple concurrent factors. Hepatology 2000;31(3):587–591.
12. Menon KV, Shah V, Kamath PS. The Budd-Chiari syndrome. N Engl J Med 2004;350(6):578–585.
13. Mitchell MC, Boitnott JK, Kaufman S, Cameron JL, Maddrey WC. Budd-Chiari syndrome: etiology, diagnosis and management. Medicine (Baltimore) 1982;61(4):199–218.
14. Walker ID. Thrombophilia in pregnancy. J Clin Pathol 2000;53(8):573–580.
15. Valla D, Casadevall N, Lacombe C, et al. Primary myeloproliferative disorder and hepatic vein thrombosis: a prospective study of erythroid colony formation in vitro in 20 patients with Budd-Chiari syndrome. Ann Intern Med 1985;103(3):329–334.
16. Espinosa G, Font J, García-Pagan JC, et al. Budd-Chiari syndrome secondary to antiphospholipid syndrome: clinical and immunologic characteristics of 43 patients. Medicine (Baltimore) 2001;80(6): 345–354.
17. Levine JS, Branch DW, Rauch J. The antiphospholipid syndrome. N Engl J Med 2002;346(10): 752–763.
18. Mohanty D, Shetty S, Ghosh K, Pawar A, Abraham P. Hereditary thrombophilia as a cause of Budd-Chiari syndrome: a study from Western India. Hepatology 2001;34(4 pt 1):666–670.
19. Mahmoud AE, Mendoza A, Meshikhes AN, et al. Clinical spectrum, investigations and treatment of Budd-Chiari syndrome. QJM 1996;89(1):37–43.
20. Hirshberg B, Shouval D, Fibach E, Friedman G, Ben-Yehuda D. Flow cytometric analysis of autonomous growth of erythroid precursors in liquid culture detects occult polycythemia vera in the Budd-Chiari syndrome. J Hepatol 2000;32(4): 574–578.
21. Girolami A, Simioni P, Girolami B, Radossi P. Homozygous patients with APC resistance may remain paucisymptomatic or asymptomatic during oral contraception. Blood Coagul Fibrinolysis 1996;7(6): 590–594.
22. Valla D, Le MG, Poynard T, Zucman N, Rueff B, Benhamou JP. Risk of hepatic vein thrombosis in relation to recent use of oral contraceptives: a case-control study. Gastroenterology 1986;90(4): 807–811.
23. Valla DC. The diagnosis and management of the Budd-Chiari syndrome: consensus and controversies. Hepatology 2003;38(4):793–803.
24. Rosenberg RD, Aird WC. Vascular-bed–specific hemostasis and hypercoagulable states. N Engl J Med 1999;340(20):1555–1564.
25. Moreira V, Aller R, De Luis DA, San Roman AL, Ferreiro A. Fulminant acute Budd-Chiari syndrome stemming from an adrenal tumor. J Clin Gastroenterol 1997;24(2):110–112.
26. Ciancio G, Soloway M. Renal cell carcinoma invading the hepatic veins. Cancer 2001;92(7):1836–1842.
27. Fortner JG, Kallum BO, Kim DK. Surgical management of hepatic vein occlusion by tumor: Budd-Chiari syndrome. Arch Surg 1977;112(6):727–728.
28. Ku CF, Lo CY, Chan WF, Chiu SW, Fan ST, Lam KS. Resection of phaeochromocytoma extending into the right atrium in a patient with multiple endocrine neoplasia type 2A. Hong Kong Med J 2005; 11(1):59–62.
29. Xu PQ, Dang XW. Treatment of membranous Budd-Chiari syndrome: analysis of 480 cases. Hepatobiliary Pancreat Dis Int 2004;3(1):73–76.
30. Valla D, Benhamou JP. Obstruction of the hepatic veins or suprahepatic inferior vena cava. Dig Dis 1996;14(2):99–118.
31. Mori H, Hayashi K, Amamoto Y. Membranous obstruction of the inferior vena cava associated with intrahepatic portosystemic shunt. Cardiovasc Intervent Radiol 1986;9(4):209–213.
32. Slakey DP, Klein AS, Venbrux AC, Cameron JL. Budd-Chiari syndrome: current management options. Ann Surg 2001;233(4):522–527.
33. Ganguli SC, Ramzan NN, McKusick MA, Andrews JC, Phyliky RL, Kamath PS. Budd-Chiari syndrome in patients with hematological disease: a therapeutic challenge. Hepatology 1998;27(4):1157–1161.
34. Klein AS, Cameron JL. Diagnosis and management of the Budd-Chiari syndrome. Am J Surg 1990;160(1):128–133.
35. Orloff MJ, Daily PO, Orloff SL, Girard B, Orloff MS. A 27-year experience with surgical treatment of Budd-Chiari syndrome. Ann Surg 2000;232(3): 340–352.
36. Langlet P, Escolano S, Valla D, et al. Clinicopathological forms and prognostic index in Budd-Chiari syndrome. J Hepatol 2003;39(4):496–501.
37. McCuskey RS, Reilly FD. Hepatic microvasculature: dynamic structure and its regulation. Semin Liver Dis 1993;13(1):1–12.
38. McCuskey RS. Morphological mechanisms for regulating blood flow through hepatic sinusoids. Liver 2000;20(1):3–7.
39. Noone TC, Semelka RC, Siegelman ES, et al. Budd-Chiari syndrome: spectrum of appearances of acute, subacute, and chronic disease with magnetic resonance imaging. J Magn Reson Imaging 2000;11 (1):44–50.
40. Tang TJ, Batts KP, de Groen PC, et al. The prognostic value of histology in the assessment of patients with Budd-Chiari syndrome. J Hepatol 2001;35(3): 338–343.
41. Henderson JM, Warren WD, Millikan WJ Jr, et al. Surgical options, hematologic evaluation, and pathologic changes in Budd-Chiari syndrome. Am J Surg 1990;159(1):41–48; discussion 48–50.

42. Tanaka M, Wanless IR. Pathology of the liver in Budd-Chiari syndrome: portal vein thrombosis and the histogenesis of veno-centric cirrhosis, veno-portal cirrhosis, and large regenerative nodules. Hepatology 1998;27(2):488–496.
43. Klein AS, Molmenti EP. Surgical treatment of Budd-Chiari syndrome. Liver Transpl 2003;9(9): 891–896.
44. Kondo F. Benign nodular hepatocellular lesions caused by abnormal hepatic circulation: etiological analysis and introduction of a new concept. J Gastroenterol Hepatol 2001;16(12):1319–1328.
45. Cazals-Hatem D, Vilgrain V, Genin P, et al. Arterial and portal circulation and parenchymal changes in Budd-Chiari syndrome: a study in 17 explanted livers. Hepatology 2003;37(3):510–519.
46. Kane R, Eustace S. Diagnosis of Budd-Chiari syndrome: comparison between sonography and MR angiography. Radiology 1995;195(1):117–121.
47. Ralls PW, Johnson MB, Radin DR, Boswell WD Jr, Lee KP, Halls JM. Budd-Chiari syndrome: detection with color Doppler sonography. AJR Am J Roentgenol 1992;159(1):113–116.
48. Zwiebel WJ. Sonographic diagnosis of hepatic vascular disorders. Semin Ultrasound CT MR 1995; 16(1):34–48.
49. Soyer P, Rabenandrasana A, Barge J, et al. MRI of Budd-Chiari syndrome. Abdom Imaging 1994; 19(4):325–329.
50. Noone TC, Semelka RC, Woosley JT, Pisano ED. Ultrasound and MR findings in acute Budd-Chiari syndrome with histopathologic correlation. J Comput Assist Tomogr 1996;20(5):819–822.
51. Mathieu D, Vasile N, Menu Y, Van Beers B, Lorphelin JM, Pringot J. Budd-Chiari syndrome: dynamic CT. Radiology 1987;165(2):409–413.
52. Miller WJ, Federle MP, Straub WH, Davis PL. Budd-Chiari syndrome: imaging with pathologic correlation. Abdom Imaging 1993;18(4):329–335.
53. Mitchell DG, Nazarian LN. Hepatic vascular diseases: CT and MRI. Semin Ultrasound CT MR 1995;16(1):49–68.
54. Stanley P. Budd-Chiari syndrome. Radiology 1989; 170(3 pt 1):625–627.
55. Vilgrain V, Lewin M, Vons C, et al. Hepatic nodules in Budd-Chiari syndrome: imaging features. Radiology 1999;210(2):443–450.
56. Brancatelli G, Federle MP, Grazioli L, Golfieri R, Lencioni R. Large regenerative nodules in Budd-Chiari syndrome and other vascular disorders of the liver: CT and MR imaging findings with clinicopathologic correlation. AJR Am J Roentgenol 2002; 178(4):877–883.
57. Janssen HL, Garcia-Pagan JC, Elias E, et al. Budd-Chiari syndrome: a review by an expert panel. J Hepatol 2003;38(3):364–371.
58. Murad SD, Valla DC, de Groen PC, et al. Determinants of survival and the effect of portosystemic shunting in patients with Budd-Chiari syndrome. Hepatology 2004;39(2):500–508.
59. Arroyo V, Ginès P, Gerbes AL, et al. Definition and diagnostic criteria of refractory ascites and hepatorenal syndrome in cirrhosis. International Ascites Club. Hepatology 1996;23(1):164–176.
60. Lang H, Oldhafer KJ, Kupsch E, Ringe B, Pichlmayr R. Liver transplantation for Budd-Chiari syndrome—palliation or cure? Transpl Int 1994;7(2): 115–119.
61. Fisher NC, McCafferty I, Dolapci M, et al. Managing Budd-Chiari syndrome: a retrospective review of percutaneous hepatic vein angioplasty and surgical shunting. Gut 1999;44(4):568–574.
62. Cooper S, Olliff SP, Elias E. Recanalisation of hepatic veins by a combined transhepatic, transjugular approach in three cases of Budd Chiari syndrome. J Interv Radiol 1996;11:9–13.
63. Sharma S, Texeira A, Texeira P, Elias E, Wilde J, Olliff SP. Pharmacological thrombolysis in Budd Chiari syndrome: a single centre experience and review of the literature. J Hepatol 2004;40(1):172–180.
64. Haskal ZJ. Use of the Arrow percutaneous thrombectomy device and transjugular liver biopsy cannula for mechanical thrombectomy of the inferior vena cava in a patient with Budd-Chiari syndrome. J Vasc Interv Radiol 2007;18(7):924–927.
65. Michl P, Bilzer M, Waggershauser T, et al. Successful treatment of chronic Budd-Chiari syndrome with a transjugular intrahepatic portosystemic shunt. J Hepatol 2000;32(3):516–520.
66. Ochs A, Sellinger M, Haag K, et al. Transjugular intrahepatic portosystemic stent-shunt (TIPS) in the treatment of Budd-Chiari syndrome. J Hepatol 1993;18(2):217–225.
67. Ryu RK, Durham JD, Krysl J, et al. Role of TIPS as a bridge to hepatic transplantation in Budd-Chiari syndrome. J Vasc Interv Radiol 1999;10(6): 799–805.
68. Brems JJ, Hiatt JR, Klein AS, et al. Effect of a prior portasystemic shunt on subsequent liver transplantation. Ann Surg 1989;209(1):51–56.
69. Cejna M, Peck-Radosavljevic M, Schoder M, et al. Repeat interventions for maintenance of transjugular intrahepatic portosystemic shunt function in patients with Budd-Chiari syndrome. J Vasc Interv Radiol 2002;13(2 pt 1):193–199.
70. Kuo PC, Johnson LB, Hastings G, et al. Fulminant hepatic failure from the Budd-Chiari syndrome: a bridge to transplantation with transjugular intrahepatic portosystemic shunt. Transplantation 1996; 62(2):294–296.
71. Shrestha R, Durham JD, Wachs M, et al. Use of transjugular intrahepatic portosystemic shunt as a bridge to transplantation in fulminant hepatic failure due to Budd-Chiari syndrome. Am J Gastroenterol 1997;92(12):2304–2306.
72. Hernández-Guerra M, Turnes J, Rubinstein P, et al. PTFE-covered stents improve TIPS patency in Budd-Chiari syndrome. Hepatology 2004;40(5): 1197–1202.
73. Gandini R, Konda D, Simonetti G. Transjugular intrahepatic portosystemic shunt patency and clinical outcome in patients with Budd-Chiari syndrome: covered versus uncovered stents. Radiology 2006; 241(1):298–305.

This article meets the criteria for 1.0 credit hour in category 1 of the AMA Physician's Recognition Award. To obtain credit, see accompanying test at http://www.rsna.org/education/rg_cme.html.

RadioGraphics

Diagnostic and Interventional Radiology for Budd-Chiari Syndrome

Marco Cura, MD, et al

RadioGraphics 2009; 29:669–681 • **Published online** 10.1148/rg.293085056 • **Content Codes:** GI VI

Page 671
Inability of blood flow to drain from the liver leads to increased sinusoidal pressure and hepatic congestion. The obstruction of hepatopetal portal venous flow causes portal hypertension. The ensuing venous stasis increases the sinusoidal pressure, leading to a reduction of portal perfusion.

Page 673
Consecutive liver biopsies in patients with Budd-Chiari syndrome are useful for assessing the severity of disease (congestion, necrosis, fibrosis, and regenerative nodules) and determining whether it has progressed after venous decompression procedures. Because of the inhomogeneous distribution of disease in the liver, a single biopsy result is less significant than the trend of histopathologic changes seen in consecutive liver biopsy specimens. Specimens obtained after adequate treatment (treatment that restores sufficient liver venous outflow) may exhibit improvement in, or total resolution of, liver congestion.

Page 674
IVC stenosis and occlusion secondary to liver congestion and compensatory caudate lobe hypertrophy are common in Budd-Chiari syndrome.

Page 674
The primary goal of treatment is the resolution of hepatic congestion in order to improve liver perfusion and preserve functioning hepatocytes. The available therapies in Budd-Chiari syndrome are diverse, and factors that influence the prognosis should be considered when stratifying risks and selecting a therapy. The selection of a specific treatment method is directed by whether the predominant manifestation is liver failure or portal hypertension. It also depends on the potential for parenchymal recovery, the surgical risk, and the availability of a liver donor.

Page 679
In patients in whom the hepatic vein cannot be recanalized or in whom recanalization produces inadequate liver decongestion, TIPS creation is a good alternative method for relieving liver congestion, with lower mortality and morbidity than those associated with open surgical procedures. ... Not only is the procedure less invasive than open surgery, but a TIPS also has the advantage over side-to-side portocaval, mesocaval, and splenorenal portosystemic shunts because it bypasses the caval stenosis. Like a mesoatrial shunt, a TIPS provides venous decompression by allowing outflow into the suprahepatic IVC, at a site proximal to the intrahepatic stenosis.