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Interventional Radiology and Surgical Treatment Options for Non-Cirrhotic Portal Hypertension

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Abstract

Purposeof review Noncirrhotic portal hypertension (NCPH) consists of a heterogenous group of disorders that lead to portal hypertension (pHTN) in the absence of cirrhosis and can be best understood by their relationship to hepatic vascular anatomy. Here, we discuss the most common of these: portal vein thrombosis, porto-sinusoidal vascular disease, Budd-Chiari syndrome, and hepatic sinusoidal obstruction syndrome.

Recent advances We provide a brief overview of each disorder and highlight recent advances in interventional radiology and surgical treatments. Although the former have improved outcomes, the latter may still be indicated in specific situations.

Summary NCPH requires careful diagnostic and therapeutic evaluation. Treatments are evolving to be less invasive, leading to improved outcomes.

Keywords Portal hypertension · Portal vein thrombosis · Porto-sinusoidal vascular disorder · Budd-Chiari syndrome · Sinusoidal obstruction syndrome

Introduction

Portal hypertension (pHTN) refers to a syndrome of increased pressure within the portomesenteric system. In the Western world, it is most commonly caused by cirrhosis, which is associated with a hepatic venous pressure gradient of > 5 mmHg. By contrast, non-cirrhotic portal hypertension (NCPH) consists of a heterogenous group of predominantly vascular disorders that produce pHTN in the absence of cirrhosis [1]. In general, the causes can be divided based on the site of pathology into pre-hepatic, hepatic, and post-hepatic, with the hepatic category further subdivided into pre-sinusoidal, sinusoidal, and post-sinusoidal (Fig. 1). In this review, we discuss the advances in interventional radiology and surgical treatments for four of these disease entities: portal vein thrombosis (PVT), porto-sinusoidal vascular disease (PSVD), Budd-Chiari syndrome (BCS), and hepatic sinusoidal obstruction syndrome (SOS), previously known as veno-occlusive disease.

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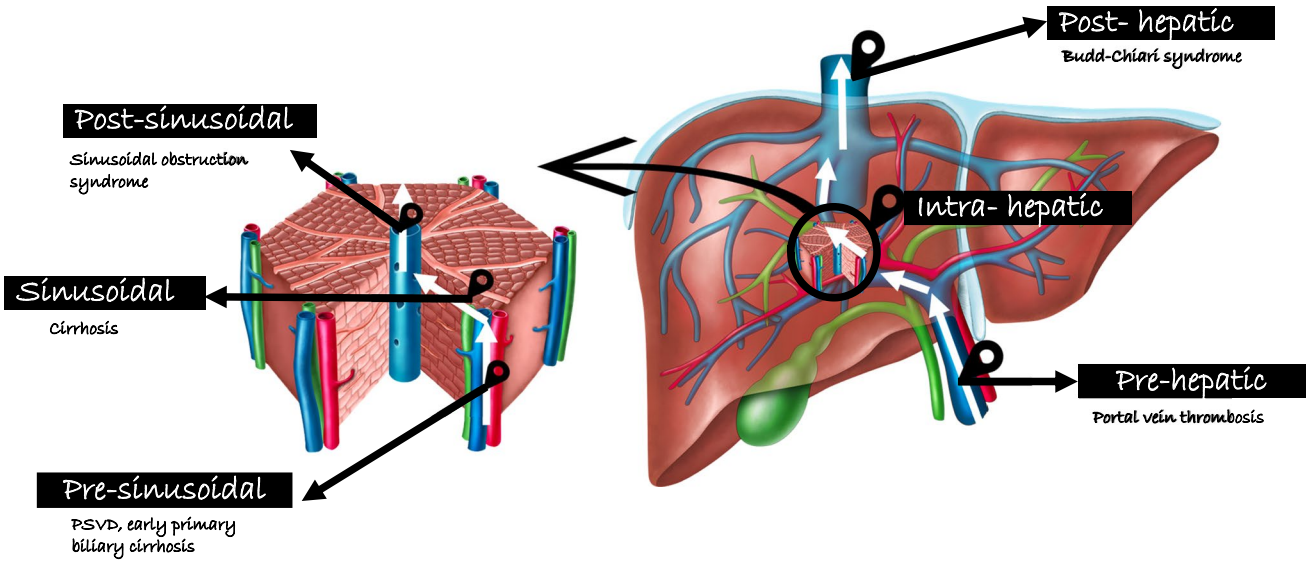

Fig. 1 Categories and common etiologies of portal hypertension. An increase in pressure in any part of this system leads to portal hypertension

Portal Vein Thrombosis

PVT is the most common etiology of pre-hepatic pHTN. It may be caused by inherited or acquired prothrombotic disorders, myeloproliferative neoplasm (MPN), portal vein invasion by local malignancy, or regional inflammation (e.g., pancreatitis, inflammatory bowel disease). The main complications are variceal bleeding and bowel ischemia due to venous congestion and the resultant low-flow state. The treatment of PVT depends on its acuity, although no precise cutoff exists to differentiate acute from chronic [1].

Acute Portal Vein Thrombosis

Management

The first-line treatment for acute PVT is anticoagulation, with the goal of both preventing clot extension to avoid intestinal infarction and achieving portal vein recanalization to prevent the eventual development of chronic pHTN [2]. In a European study of 95 patients with non-cirrhotic nonmalignant PVT who were treated with early administration of low molecular weight heparin and then transitioned to warfarin, no patients experienced thrombus extension or death, although intestinal infarction occurred in two. Complete portal vein recanalization was seen in only one-third of patients after 6 months of treatment, and 40% developed cavernous transformation at one year [3].

Treatment of the underlying etiology of thrombosis is also important. For example, cytoreductive therapy for MPN has been shown to improve outcomes, and resolution of pancreatitis often removes the nidus for clot formation [2]. If the etiology is unknown, this should prompt diagnostic investigation as spontaneous PVT is rare.

Transcatheter thrombolytic administration via transsplenic or transhepatic access is rarely used in the acute setting given high complication rates and similar rates of recanalization compared with systemic anticoagulation. More recently, it was shown that a combination of transjugular thrombectomy and local fibrinolysis with or without transjugular intrahepatic portosystemic shunt (TIPS) placement achieved a 94% recanalization rate in patients with non-cirrhotic nonmalignant acute PVT at imminent risk of intestinal infarction. Patency was maintained at 88% at 2 years, and major non-fatal complications were observed in 3 of 17 patients [4]. Consequently, this approach should be considered in patients with evidence of intestinal ischemia.

The role of surgery is limited in acute PVT except in the setting of intestinal infarction. Unlike mesenteric ischemia resulting from arterial inflow obstruction, ischemia due to occlusion of venous outflow is less abrupt in onset. It results in progressive bowel congestion, ultimately causing infarction when the backpressure overwhelms and inhibits the arterial inflow. This manifests as discomfort, nausea/emesis, or both for a least two days prior to presentation. When diagnosed, conservative management with bowel rest and serial abdominal examinations should be the initial treatment, with

catheter-directed therapy selectively employed as described above.

Suspected intestinal necrosis should prompt consideration of surgical exploration. In unclear cases, diagnostic laparoscopy can be considered. However, the profound bowel edema often precludes the creation of a clear working space and pneumoperitoneum can worsen the already-diminished mesenteric blood flow [5]. As such, open exploration is generally preferred. Also, determining the margins between infarcted and viable bowel is particularly challenging as the boundaries are poorly defined, and adjunctive techniques to assess viability, such as fluorescein and Doppler flow assessment, are less useful [6]. Consequently, temporary abdominal closure and re-exploration should be liberally employed, which has the added benefit of reducing the risk of abdominal compartment syndrome [7].

The distribution of ischemia is related to the extent of thrombosis. In cases of isolated portomesenteric occlusion, ischemia is usually limited to the small bowel [8]. However, if the splenic and inferior mesenteric veins are occluded, colonic infarction may also occur. In cases of pan-intestinal ischemia, only the grossly non-viable or perforated bowel should be resected initially, with re-exploration performed after initiation of anticoagulation therapy. Surgical thrombectomy alone is rarely indicated but can be undertaken if laparotomy is performed for bowel ischemia [9]. This is typically performed via venotomy in the superior mesenteric vein (SMV) where it can be easily controlled in the lesser sac just inferior to the pancreas using a Fogarty balloon. Postoperative catheter-directed thrombolysis performed by infusing papaverine with or without a thrombolytic agent into the superior mesenteric *artery* has also been reported. In one clinical trial, compared to systemic anticoagulation alone, this technique improved thrombus resolution, need for additional bowel resection, and survival [10].

Chronic Portal Vein Thrombosis

Management

In contrast to acute PVT, chronic PVT will not resolve with anticoagulation alone. In patients with recurrent and frequent complications of pHTN, TIPS placement with or without portal vein recanalization should be considered. Importantly, it may improve both liver function and transplant candidacy. However, the procedure may be technically challenging in cases of a diminutive or cavernous portal vein and frequently requires additional access through the spleen, liver, or superior mesenteric vein which can increase procedure time and bleeding risk. In rare cases, a mesocaval shunt bridging the inferior vena cava (IVC) and a residual vein in the portomesenteric system may be necessary, although the performance of this procedure requires specialized expertise.

Evidence in support of portal vein recanalization combined with TIPS (PVR-TIPS) is predominantly from patients with cirrhosis awaiting liver transplantation, in whom the procedure has shown high rates of success. From a technical perspective, the utilization of trans-splenic access is superior to a transhepatic approach, resulting in a 100% technical success rate and fewer adverse effects [11]. This secondary access allows identification of the diminutive portal vein using the left gastric vein as a marker that, once cannulated and dilated, can act as a target for transhepatic needle passage. In patients with neither cirrhosis nor sinusoidal pHTN, TIPS might be unnecessary if the distal intrahepatic portal vein branches are not occluded [12, 13]. A recent study showed that in chronic non-cirrhotic extrahepatic portal vein occlusion (EHPVO), portal vein recanalization without TIPS insertion was feasible and safe [14, 15].

With the proliferation of catheter-based therapy and increasingly effective medical or endoscopic treatment to address the sequelae of chronic non-cirrhotic EHPVO, the need for surgery in these patients has been greatly reduced. It is limited to patients with intractable complications (usually bleeding) not amenable to the approaches described above. Surgical options are dictated by patient anatomy and the extent of venous occlusion. In cases where both the SMV and left portal vein are patent, the meso-Rex bypass is preferred as it re-establishes flow via the native portal system. So named as the distal shunt target is the recessus of Rex in the umbilical fissure of the left liver, the meso-Rex bypass was popularized in the pediatric transplant population for EHPVO but can be utilized in adults [16–18]. Unfortunately, the SMV is often involved in adult EHPVO. In the uncommon case where the SMV and PV are thrombosed but the splenic vein remains patent, a splenorenal (Warren-type) shunt can be performed with good results. In cases with long-segment occlusion of the PV, SMV, and splenic vein, multivisceral transplantation is sometimes necessary [17].

Porto-Sinusoidal Vascular Disorder

Porto-sinusoidal vascular disorder (PSVD) was first proposed by the Vascular Liver Diseases Interest Group (VALDIG) in 2017 to describe a heterogenous group of conditions causing non-cirrhotic pHTN that mainly involves the portal venules or pre-sinusoidal areas [19].

Causes of PSVD include medications, toxins, immunologic disorders, genetic conditions, and prothrombotic states (Table 1). The presence of other liver diseases such as viral hepatitis, alcohol use disorder, metabolic syndrome, or PVT does not exclude the diagnosis of PSVD as they can coexist [20].

Table 1 Different mechanisms of injury leading to PSVD

Categories	Examples
Medications/Toxins	Thiopurine derivatives (e.g., azathioprine)
	Oxaliplatin
	Arsenicals / vinylchloride
Immunological disorders	Common variable immunodeficiency syndrome (CVID)
	Hyper-IgM syndrome
	Connective tissue diseases (SLE, scleroderma, RA)
	Celiac disease
	Autoimmune hepatitis
	Primary antibody-deficiency syndromes (Bruton, Felty)
	Solid organ & hematopoietic progenitor cell transplantation
Infections	Intra-abdominal infections
	HIV infection
Genetic disorders	Cystic fibrosis
	Polycystic liver disease (PKD-1 mutation)
	Turner's syndrome
	Familial obliterative portal venopathy
Prothrombotic conditions	Inherited thrombophilias Factor V Leiden mutation, protein C or S or antithrombin deficiency
	Acquired thrombophilias Antiphospholipid syndrome, myeloproliferative neoplasms

Specific diagnostic criteria have been established for the diagnosis of PSVD. Cirrhosis must first be excluded via liver biopsy. Following this, the diagnosis can be entertained if one of the following is present: [1] one specific sign of portal hypertension (varices, portosystemic collaterals), [21] one specific histological lesion (obliterative portal venopathy, nodular regenerative hyperplasia, or incomplete septal fibrosis), or [2] both one non-specific sign of portal hypertension and one non-specific histological lesion.

Management

Once PSVD has been diagnosed, an attempt to identify the underlying etiology should ensue. Specific attention should be given to the presence of immunological, thrombophilic, and genetic disorders, as well as exposure to drugs/toxins (Table 1). Patients should undergo esophagogastroduodenoscopy (EGD) for variceal screening. In patients with features of pHTN, Doppler ultrasound should be performed every 6 months as the yearly probability of developing PVT is 9% [22]. There is no proven role for prophylactic anticoagulation and, in those who develop PVT, guidelines for PVT in patients with cirrhosis should be employed [20].

TIPS placement is useful for the management of severe complications of portal hypertension. These patients typically have excellent outcomes if they have normal kidney function and no severe extrahepatic comorbidities [23]. In fact, patients with PSVD have fewer TIPS-related complications than those with cirrhosis. In one study comparing these groups following TIPS insertion for management of variceal bleeding, patients with PSVD had significantly lower rates of hepatic encephalopathy, hepatic impairment, and long-term mortality [24].

Budd-Chiari Syndrome

BCS refers to obstruction of hepatic venous outflow, which can occur at any level from the hepatic venules to the IVC. It is divided into primary, which results from thrombosis, and secondary, which is caused by venous compression or invasion, typically by malignancy. Primary BCS occurs most commonly in prothrombotic states so, when diagnosed, the existence/type of clotting disorder should be elucidated.

Management

Anticoagulation should be initiated in all patients regardless of symptomatology to achieve recanalization and prevent thrombosis progression [2]. However, even in those started on anticoagulation at diagnosis, the 5-year intervention-free survival rate is only 30%. The underlying disease leading to the development of BCS, such as MPN or paroxysmal nocturnal hemoglobinuria, should be identified and treated. Where present, complications of pHTN should be managed similarly to when they occur in the setting of cirrhosis.

Catheter-directed thrombolysis involves the percutaneous insertion of a catheter into a hepatic vein and the instillation

of thrombolytics over several hours. Small series have reported promising results, particularly in patients with acute and incomplete thrombosis and when combined with other interventions, such as angioplasty or stent placement [25]. However, limited evidence and the risk of hemorrhage have precluded widespread adoption.

Percutaneous angioplasty with or without stent placement is an effective approach for restoring the physiological hepatic outflow, especially in the setting of segmental stenosis or suprahepatic IVC obstruction. It is important to identify these patients as soon as the diagnosis of BCS is made. Very high success rates have been reported among Asian cohorts with combined angioplasty and stenting, resulting in a cumulative 5-year primary patency rate of 77% [26]. The success rate of angioplasty is lower in comparable European cohorts, with nearly 64% of patients requiring re-intervention (TIPS or transplant) during 5-year follow-up [27]. This difference in outcomes is likely related to the underlying etiology of BCS in these patients; venous stenosis, which is particularly responsive to this form of treatment, was present in the majority of patients in the Asian cohort while it was present in only 10% of the European cohort [27].

Flow diversion procedures utilize a shunt to redirect portal flow past the obstruction, most commonly in the form of a TIPS. Importantly, this should not be performed in patients whose site of obstruction is downstream of the hepatic veins, as the shunt would be of no benefit since it would not bypass the obstruction. Where indicated, TIPS placement has demonstrated improved transplant-free survival and lower morbidity and mortality compared with shunt surgery [28, 29]. Although repeated endovascular revisions were needed in 42% of patients, the rate of secondary patency was close to 100%, and 10-year survival was 76% [30]. Of note, TIPS placement can be technically challenging due to the inaccessibility of the hepatic veins, and a transcaval approach (direct intrahepatic portosystemic shunt, or DIPS) may be necessary for up to 60% of patients. DIPS should only be performed by experienced operators at high-volume centers [29]. Surgical shunts can also achieve hepatic decompression but have largely been superseded by TIPS. Thrombectomy alone is often not technically possible. Caval patency is a prerequisite for effective portacaval, mesocaval, or splenorenal shunt construction, although mesoatrial or meso-caval-atrial shunts have been described [31, 32]. In general, shunt employment should be rare in the modern era for BCS; in particular, portacaval or mesocaval shunts can greatly complicate surgery if transplantation ensues and should be avoided.

Liver transplantation is the treatment of last resort in patients with BCS as it presents both surgical and medical challenges. Surgically, hepatomegaly and hepatic vein thrombosis increase technical complexity, particularly with vascular anastomoses. For example, enlargement of the caudate lobe and occlusion of the hepatic vein ostia make the "piggyback" technique challenging. The primary medical challenge is the underlying prothrombotic disorder and risk of recurrent thrombosis post-transplant. While inherited conditions such as factor V Leiden are cured with liver transplantation, acquired disorders such as MPN and paroxysmal nocturnal hemoglobinuria are not and place the patient at increased risk of recurrent thrombosis post-transplant. Importantly, MPN is not a contraindication to transplantation and, with appropriate anticoagulation, aspirin, and antiproliferative medications (e.g., hydroxyurea), survival rates are similar to those without MPN [33, 34].

Sinusoidal Obstruction Syndrome

SOS, also known as veno-occlusive disease (VOD), is a post-sinusoidal obstruction to venous outflow that mostly occurs following hematopoietic stem cell transplantation. The primary site of injury is sinusoidal endothelial cells and hepatocytes in zone 3 of the hepatic acinus. Mortality has been reported to be as high as 80% in severe cases [35]. The incidence of SOS varies substantially from 2 to 60% due to differences in hematopoietic stem cell transplant regimens and diagnostic criteria.

Management

SOS is classified as mild, moderate, or severe based on the timing of presentation, degree of hepatic dysfunction, and severity of volume overload. Most patients have mild SOS, which is self-limiting and requires only supportive care. However, in those with moderate or severe disease, treatment with defibrotide should be considered. Its mechanism of action is not fully understood, but it appears to act as an antithrombotic and profibrinolytic drug that reduces platelet adhesion and activation and decreases vascular permeability. It has been shown to improve 100-day overall survival in patients with SOS [36–38].

For patients with very severe and rapidly progressive SOS, early TIPS insertion has been tried in small series with inconclusive results. In older series, despite the absence of procedural complications, mortality remained high [39]. However, in a recent single-center experience of patients with very severe SOS, TIPS showed more promising results. The series included seven patients with rapid clinical deterioration despite treatment with defibrotide. The procedure was performed a median of four days after SOS diagnosis and resulted in a drop in pressure gradient from a median of 24 to 7 mmHg. Following TIPS insertion, all patients showed clinical improvement in the degree of ascites accumulation, renal failure, and liver synthetic function, and 100-day survival was 100% [40]. Although incompletely

understood, these improved outcomes may in part be related to co-treatment with defibrotide, which was not previously available.

Liver transplantation has shown generally poor outcomes, particularly in adults, with a survival rate of only 20% (3 out of 15 adult cases survived from 9 months to 8 years) [41].

Conclusions

In NCPH, the site of the blockage can be pre-hepatic, intrahepatic, or post-hepatic. Diagnosis of NCPH requires a high degree of suspicion and an understanding of the vascular anatomy of the liver. The approach to treating portal hypertensive complications in NCPH is similar to the management of complications of cirrhosis-related pHTN. Interventional radiology options for the treatment of NCPH are less invasive and have lower morbidity than most surgical interventions. However, surgery is still required in rare, severe cases of NCPH. Experienced, tertiary care teams have the best outcomes.

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Declarations

Conflict of Interest The authors declare no conflicts of interest.

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References

1. Hernández-Gea V, Baiges A, Turon F, Garcia-Pagán JC. Idiopathic Portal Pypertens Hepatol. 2018;68(6):2413–23.
2. Hernández-Gea V, De Gottardi A, Leebeek FWG, Rautou PE, Salem R, Garcia-Pagan JC. Current knowledge in pathophysiology and management of Budd-Chiari syndrome and non-cirrhotic non-tumoral splanchnic vein thrombosis. J Hepatol. 2019;71(1):175–99.
3. Plessier A, Darwish-Murad S, Hernandez-Guerra M, Consigny Y, Fabris F, Trebicka J, et al. Acute portal vein thrombosis unrelated to cirrhosis: a prospective multicenter follow-up study. Hepatology. 2010;51(1):210–8.
4. Klinger C, Riecken B, Schmidt A, De Gottardi A, Meier B, Bosch J, et al. Transjugular local thrombolysis with/without TIPS in patients with acute non-cirrhotic, non-malignant portal vein thrombosis. Dig Liver Dis. 2017;49(12):1345–52.
5. Boley SJ, Kaleya RN, Brandt LJ. Mesenteric venous thrombosis. Surg Clin North Am. 1992;72(1):183–201.
6. Urbanavičius L, Pattyn P, de Putte DV, Venskutonis D. How to assess intestinal viability during surgery: A review of techniques. World J Gastrointest Surg. 2011;3(5):59–69.
7. Khodadadi J, Rozencwajg J, Nacasch N, Schmidt B, Feuchtwanger MM. Mesenteric vein thrombosis. The importance of a second-look operation. Arch Surg. 1980;115(3):315–7.
8. Johnson CC, Baggenstoss AH. Mesenteric vascular occlusion; study of 99 cases of occlusion of veins. Proc Staff Meet Mayo Clin. 1949;24(25):628–36.
9. EASL Clinical Practice Guidelines. Vascular diseases of the liver. J Hepatol. 2016;64(1):179–202.
10. Yang S, Zhang L, Liu K, Fan X, Ding W, He C, et al. Postoperative catheter-directed thrombolysis versus systemic anticoagulation for acute superior mesenteric venous thrombosis. Ann Vasc Surg. 2016;35:88–97.
11. Thornburg B, Desai K, Hickey R, Hohlastos E, Kulik L, Ganger D, et al. Pretransplantation portal vein recanalization and transjugular intrahepatic portosystemic shunt creation for chronic portal vein thrombosis: final analysis of a 61-patient cohort. J Vasc Interv Radiol. 2017;28(12):1714-21.e2.
12. Kallini JR, Gabr A, Kulik L, Ganger D, Lewandowski R, Thornburg B, et al. Noncirrhotic complete obliterative portal vein thrombosis: novel management using trans-splenic transjugular intrahepatic portosystemic shunt with portal vein recanalization. Hepatology. 2016;63(4):1387–90.
13. Knight GM, Clark J, Boike JR, Maddur H, Ganger DR, Talwar A, et al. TIPS for adults without cirrhosis with chronic mesenteric venous thrombosis and EHPVO refractory to standard-of-care therapy. Hepatology. 2021;74(5):2735–44.
14. Artru F, Vietti-Violi N, Sempoux C, Vieira Barbosa J, Becce F, Sah N, et al. Portal vein recanalisation alone to treat severe portal hypertension in non-cirrhotic patients with chronic extrahepatic portal vein obstruction. JHEP Reports. 2022;4(8).
15. Marot A, Barbosa JV, Duran R, Deltenre P, Denys A. Percutaneous portal vein recanalization using self-expandable nitinol stents in patients with non-cirrhotic non-tumoral portal vein occlusion. Diagn Interv Imaging. 2019;100(3):147–56.
16. Superina R, Shneider B, Emre S, Sarin S, de VilledeGoyet J. Surgical guidelines for the management of extra-hepatic portal vein obstruction. Pediatr Transplant. 2006;10(8):908–13.
17. Brichard M, Iesari S, Lerut J, Reding R, Goffette P, Coubeau L. Meso-Rex bypass for the management of extrahepatic portal vein obstruction in adults (with video). Hepatobiliary Pancreat Dis Int. 2022;21(1):25–32.
18. di Francesco F, Grimaldi C, de VilledeGoyet J. Meso-Rex bypass–a procedure to cure prehepatic portal hypertension: the insight and the inside. J Am Coll Surg. 2014;218(2):23–36.
19. De Gottardi A, Rautou PE, Schouten J, Rubbia-Brandt L, Leebeek F, Trebicka J, et al. Porto-sinusoidal vascular disease: proposal and description of a novel entity. Lancet Gastroenterol Hepatol. 2019;4(5):399–411.
20. de Franchis R, Bosch J, Garcia-Tsao G, Reiberger T, Ripoll C, Abraldes JG, et al. Baveno VII – Renewing consensus in portal hypertension. J Hepatol. 2022;76(4):959–74.
21. Northup PG, Garcia-Pagan JC, Garcia-Tsao G, Intagliata NM, Superina RA, Roberts LN, 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(1):366–413.
22. Siramolpiwat S, Seijo S, Miquel R, Berzigotti A, Garcia-Criado A, Darnell A, et al. Idiopathic portal hypertension: natural history and long-term outcome. Hepatology. 2014;59(6):2276–85.
23. Bissonnette J, Garcia-Pagán JC, Albillos A, Turon F, Ferreira C, Tellez L, et al. Role of the transjugular intrahepatic portosystemic shunt in the management of severe complications of portal hypertension in idiopathic noncirrhotic portal hypertension. Hepatology. 2016;64(1):224–31.

24. Lv Y, Li K, He C, Luo B, Zhang B, Liu H, et al. TIPSS for variceal bleeding in patients with idiopathic non-cirrhotic portal hypertension: comparison with patients who have cirrhosis. Aliment Pharmacol Ther. 2019;49(7):926–39.
25. 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):172–80.
26. Han G, Qi X, Zhang W, He C, Yin Z, Wang J, et al. Percutaneous recanalization for Budd-Chiari syndrome: an 11-year retrospective study on patency and survival in 177 Chinese patients from a single center. Radiology. 2013;266(2):657–67.
27. Seijo S, Plessier A, Hoekstra J, Dell'era A, Mandair D, Rifai K, et al. Good long-term outcome of Budd-Chiari syndrome with a step-wise management. Hepatology. 2013;57(5):1962–8.
28. Neumann AB, Andersen SD, Nielsen DT, Holland-Fischer P, Vilstrup H, Grønbæk H. Treatment of Budd-Chiari syndrome with a focus on transjugular intrahepatic portosystemic shunt. World J Hepatol. 2013;5(1):38–42.
29. Garcia-Pagán JC, Heydtmann M, Raffa S, Plessier A, Murad S, Fabris F, et al. TIPS for Budd-Chiari Syndrome: long-term results and prognostics factors in 124 patients. Gastroenterology. 2008;135(3):808–15.
30. Hayek G, Ronot M, Plessier A, Sibert A, Abdel-Rehim M, Zappa M, et al. Long-term outcome and analysis of dysfunction of transjugular intrahepatic portosystemic shunt placement in chronic primary Budd-Chiari syndrome. Radiology. 2017;283(1):280–92.
31. Chen H, Zhang F, Ye Y, Cheng Y, Chen Y. Long-term follow-up study and comparison of meso-atrial shunts and meso-cavo-atrial shunts for treatment of combined Budd-Chiari syndrome. J Surg Res. 2011;168(1):162–6.
32. 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):340–52.
33. Chinnakotla S, Klintmalm GB, Kim P, Tomiyama K, Klintmalm E, Davis GL, et al. Long-term follow-up of liver transplantation for Budd-Chiari syndrome with antithrombotic therapy based on the etiology. Transplantation. 2011;92(3):341–5.
34. Melear JM, Goldstein RM, Levy MF, Molmenti EP, Cooper B, Netto GJ, et al. Hematologic aspects of liver transplantation for Budd-Chiari syndrome with special reference to myeloproliferative disorders. Transplantation. 2002;74(8):1090–5.
35. Bonifazi F, Barbato F, Ravaioli F, Sessa M, Defrancesco I, Arpinati M, et al. Diagnosis and treatment of VOD/SOS after allogeneic hematopoietic stem cell transplantation. Front Immunol. 2020;11:489.
36. Corbacioglu S, Carreras E, Mohty M, Pagliuca A, Boelens JJ, Damaj G, et al. Defibrotide for the treatment of hepatic veno-occlusive disease: final results from the International Compassionate-Use Program. Biol Blood Marrow Transplant. 2016;22(10):1874–82.
37. Kernan NA, Grupp S, Smith AR, Arai S, Triplett B, Antin JH, et al. Final results from a defibrotide treatment-IND study for patients with hepatic veno-occlusive disease/sinusoidal obstruction syndrome. Br J Haematol. 2018;181(6):816–27.
38. Richardson PG, Soiffer RJ, Antin JH, Uno H, Jin Z, Kurtzberg J, et al. Defibrotide for the treatment of severe hepatic veno-occlusive disease and multiorgan failure after stem cell transplantation: a multicenter, randomized, dose-finding trial. Biol Blood Marrow Transplant. 2010;16(7):1005–17.
39. Azoulay D, Castaing D, Lemoine A, Hargreaves GM, Bismuth H. Transjugular intrahepatic portosystemic shunt (TIPS) for severe veno-occlusive disease of the liver following bone marrow transplantation. Bone Marrow Transplant. 2000;25(9):987–92.
40. Gómez-Centurión I, Bailén R, Oarbeascoa G, Muñoz C, Luque A, Boyra ME, et al. Transjugular intrahepatic portosystemic shunt for very severe veno-occlusive disease/sinusoidal obstruction syndrome (VOD/SOS) after unmanipulated haploidentical hematopoietic stem cell transplantation with post-transplantation cyclophosphamide. Biol Blood Marrow Transplant. 2020;26(11):2089–97.
41. Ichimura K, Kawamura N, Goto R, Watanabe M, Ganchiku Y, Shimamura T, et al. Living donor liver transplantation for hepatic venoocclusive disease/sinusoidal obstruction syndrome originating from hematopoietic stem cell transplantation. Case Rep Transplant. 2022;2022:8361769.