Budd-Chiari syndrome: Management

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INTRODUCTION

Budd-Chiari syndrome is defined as hepatic venous outflow tract obstruction, independent of the level or mechanism of obstruction, provided the obstruction is not due to cardiac disease, pericardial disease, or sinusoidal obstruction syndrome (veno-occlusive disease). Primary Budd-Chiari syndrome is present when there is obstruction due to a predominantly venous process (thrombosis or phlebitis), whereas secondary Budd-Chiari syndrome is present when there is compression or invasion of the hepatic veins and/or the inferior vena cava by a lesion that originates outside of the vein (eg, a malignancy).

This topic will review the treatment of Budd-Chiari syndrome. The etiology, clinical manifestations, and diagnosis of Budd-Chiari syndrome are discussed separately. (See "Etiology of the Budd-Chiari syndrome" and "Budd-Chiari syndrome: Epidemiology, clinical manifestations, and diagnosis".)

CATEGORIZATION

When patients are diagnosed with Budd-Chiari syndrome, they are categorized based on disease duration and severity [1,2]. The categories include (see "Budd-Chiari syndrome: Epidemiology, clinical manifestations, and diagnosis", section on 'Clinical manifestations'):

- Acute fulminant Budd Chiari syndrome with liver failure – Characterized by acute liver injury with elevated aminotransferases, jaundice, hepatic encephalopathy, and an elevated

prothrombin time/international normalized ratio; hepatic encephalopathy develops within eight weeks after the development of jaundice.

- Acute nonfulminant Budd-Chiari syndrome – Clinical manifestations develop rapidly (over the course of weeks), with intractable ascites and hepatic necrosis.

- Subacute Budd-Chiari syndrome – Insidious onset, with patients taking up to three months to develop symptoms. Ascites and hepatic necrosis may be minimal due to decompression of the sinusoids by portal and hepatic venous collaterals.

- Chronic Budd-Chiari syndrome – Patients present with complications of cirrhosis.

Patients with acute liver failure or acute nonfulminant liver disease have not yet developed venous collaterals, whereas venous collaterals are seen in patients with subacute and chronic liver disease.

NATURAL HISTORY AND PROGNOSIS

Symptomatic Budd-Chiari syndrome has a high mortality rate if untreated. In a series performed before specific therapy became available, 90 percent of patients died within three years [3]. Patients often die from intractable ascites with emaciation, gastrointestinal bleeding, and liver failure.

With treatment, survival rates are good. In a series of 163 patients who were treated using a stepwise approach such as that outlined below, survival rates at one, two, and five years were 87, 82, and 74 percent, respectively [4]. (See 'Management overview' below.)

However, studies have suggested that patients with Budd-Chiari syndrome have a higher risk of mortality than the general population [4-12]. In a population-based study comparing 478 patients with Budd-Chiari syndrome with over 4600 reference individuals matched for age, sex, and geographic location, patients with Budd-Chiari syndrome had higher risk of mortality (70 versus 28 deaths per 1000 person-years, adjusted hazard ratio [HR] 3.1, 95% CI 2.6-3.6) [10]. Mortality within the first year following diagnosis of Budd-Chiari syndrome was high and was linked to underlying malignant disease.

Factors associated with a worse prognosis in hospitalized patients include older age, history of cancer, hepatorenal syndrome, acute liver failure, acute respiratory failure, and ascites [12]. In addition, occlusion of the portal vein in patients with Budd-Chiari syndrome limits therapeutic options and generally has a limited prognosis [13].

MANAGEMENT

The approach to management in patients with Budd-Chiari syndrome depends on clinical and anatomic features as well as local expertise [14,15].

Goals of therapy — Treatment for Budd Chiari syndrome aims to:

- Prevent the propagation of the clot
- Restore patency of thrombosed veins
- Decompress the congested liver
- Prevent or manage complications

Management overview — We use a stepwise approach for managing patients with Budd-Chiari syndrome (algorithm 1) [16].

Initial treatment includes:

- Correcting underlying disorders that predisposed to the development of Budd-Chiari syndrome (when possible)
- Initiating anticoagulation unless there are contraindications
- Treating complications of portal hypertension (if present)

Additional treatments that may be appropriate for some patients include:

- Thrombolysis for patients with acute Budd-Chiari syndrome if a well-defined clot is present. (See 'Categorization' above.)
- Angioplasty/stenting for symptomatic patients with venous obstructions that are amenable to angiographic treatment.
- Liver transplantation for patients with acute liver failure; patients with acute liver failure should be referred to a liver transplantation center if possible for evaluation and management. (See "Acute liver failure in adults: Management and prognosis".)

If initial treatments fail, options include:

- TIPS placement or surgical shunting to decompress the liver

- Liver transplantation

In addition to treatments aimed at managing the hepatic venous outflow obstruction, patients with long-standing Budd-Chiari syndrome also need to be monitored for complications of portal hypertension and hepatocellular carcinoma, and for any underlying myeloproliferative neoplasm. Finally, patients with Budd-Chiari syndrome should undergo a nutritional evaluation, especially if there has been weight loss or other evidence of malnutrition, and attempts should be made to improve nutritional status. (See "Evaluating nutritional status in adults with cirrhosis".)

Randomized trials in patients with Budd-Chiari syndrome are lacking. Thus, the approach to management is supported by retrospective studies and clinical experience. The efficacy of stepwise therapy was studied in a series of 163 patients with Budd-Chiari syndrome treated in academic and large regional hospitals in Europe [4]. Anticoagulation was given to 140 patients (86 percent) and 80 patients were managed without undergoing invasive procedures (eg, angioplasty, thrombolysis, TIPS placement, surgical shunting, or liver transplantation). Overall survival rates were 87 and 82 percent at 12 and 24 months, respectively, and transplantation-free survival rates were 77 and 68 percent, respectively. In a follow-up study that included 157 of the original patients with a median follow-up of 50 months, overall survival rates were 79 and 74 percent at three and five years, respectively [8]. Transplantation-free survival rates were 67 and 64 percent, respectively. By comparison, historic series have suggested overall survival rates of 10 percent at three years without treatment [3]. In another study of 136 patients with Budd-Chiari Syndrome who were treated with either recanalization of the hepatic vein/inferior vena cava or creation of an intrahepatic portosystemic shunt, transplant-free survival for the entire cohort was 94 percent at one year and five years, with no significant difference in survival between the groups [17].

Treat predisposing conditions — An underlying disorder can be identified in over 80 percent of patients with Budd-Chiari syndrome (table 1) [4,18-24]. If a treatable disorder is identified, appropriate therapy should be initiated without delay [25]. (See "Etiology of the Budd-Chiari syndrome" and "Budd-Chiari syndrome: Epidemiology, clinical manifestations, and diagnosis", section on 'Evaluating for predisposing conditions'.)

Most patients have an underlying hypercoagulable disorder and require chronic anticoagulation. Exceptions include patients who develop Budd-Chiari syndrome as a result of a web in the inferior vena cava or other obstructive lesion that can be corrected surgically or radiologically [26].

The management of patients with an underlying myeloproliferative neoplasm (eg, polycythemia vera, essential thrombocythemia) is discussed separately. (See "Prognosis and treatment of polycythemia vera and secondary polycythemia" and "Prognosis and treatment of essential thrombocythemia" and "Overview of the myeloproliferative neoplasms".)

Prevent propagation of the clot — Anticoagulation should be initiated as soon as possible in most patients to prevent propagation of the clot, provided there are no contraindications. However, the risk of anticoagulation should be considered, especially in patients who present with bleeding complications or who have varices. Prior to initiating anticoagulation, we perform an upper endoscopy to screen for varices. We give anticoagulation to patients with a history of gastrointestinal variceal bleeding or varices who are at increased risk for bleeding (particularly in those with cirrhosis) only if adequate prophylactic measures to prevent recurrent bleeding can be implemented. We prefer use of a beta blocker over esophageal variceal ligation (EVL) for prophylaxis in such patients because of the risk of bleeding from esophageal ulcers that form when the bands used for EVL slough off. (See "Primary prevention of bleeding from esophageal varices in patients with cirrhosis", section on 'Preventive strategies' and "Prevention of recurrent bleeding from esophageal varices in patients with cirrhosis".)

We prefer to treat initially with low molecular weight heparin. In selected patients, such as those at high risk for bleeding or who have renal impairment, we will check anti-factor Xa activity, targeting it to between 0.5 and 0.8 international unit/mL. We also start patients on an oral vitamin K antagonist (eg, warfarin). Once the international normalized ratio is between two and three, we discontinue the low molecular weight heparin. Anticoagulation is continued indefinitely unless a major contraindication is present, a complication occurs, or the obstruction is due to an anatomic cause that has been corrected. (See "Warfarin and other VKAs: Dosing and adverse effects" and "Clinical use of coagulation tests", section on 'Monitoring heparins'.)

However, sufficient recanalization of occluded vessels or the development of adequate collateral circulation often does not occur in patients treated with anticoagulation alone [4]. The use of anticoagulation as sole therapy for the Budd-Chiari syndrome is generally reserved for:

- Patients with chronic or subacute Budd-Chiari syndrome and well-compensated liver disease at the time of presentation; however, even in these patients, additional measures to decompress the liver should be considered (particularly if they are not candidates for liver transplantation) in an attempt to delay progression of the liver disease. (See 'Categorization' above.)

- Patients in whom other types of therapy are not feasible.

Restore patency of thrombosed veins — Methods available to help restore patency of thrombosed veins include thrombolytic therapy for patients with acute Budd-Chiari syndrome (usually in patients symptomatic for only a few weeks with a well-defined clot). Treatment may also include angioplasty/stenting for selected patients with acute or subacute Budd-Chiari syndrome with segmental hepatic vein or inferior vena cava obstruction. Treatment in patients with chronic Budd-Chiari syndrome typically focuses on decompressing the liver (TIPS placement, surgical shunts, or liver transplantation), rather than restoring the patency of thrombosed veins. (See 'Decompress the liver' below.)

Thrombolytic therapy — Thrombolytic therapy is an option for treating patients with a clot that is known to be recent (less than three to four weeks old) and is well-defined on venography, provided there are no contraindications to the use of thrombolytic agents. We avoid using thrombolytic agents in patients who have an extensive clot involving the intrahepatic vena cava and hepatic veins (because of technical difficultly performing thrombolysis in such patients) or who have a clot of unknown age.

Published experience with thrombolytic agents is limited to small case series and individual reports, which have documented successful treatment [27,28]. However, enthusiasm for thrombolytic therapy in centers caring for a relatively large number of patients with Budd-Chiari syndrome is less than the published reports indicate. In most cases, treatment has limited efficacy and exposes the patient to the risks of thrombolytic therapy, which include bleeding, stroke, and pulmonary embolism.

Thrombolytic agents have been given both systemically and locally for the treatment of Budd-Chiari syndrome [27,29,30]. Although no study has directly compared the efficacy of local infusion with systemic infusion, the ability to direct infusion to an affected vein could theoretically lead to a higher concentration of the thrombolytic agent and greater efficacy. The amount of medication administered during local infusion causes a comparable coagulopathy to systemic administration; thus, the bleeding risk is similar.

Thrombolytic agents should **not** be used in the treatment of the chronic form of Budd-Chiari syndrome. Clots in such patients have matured and are unlikely to be recanalized by thrombolytic agents. Furthermore, because such patients may have developed portal hypertension, treatment can be associated with disastrous bleeding complications.

Angioplasty and stenting — Radiologically-guided treatment, including angioplasty and stenting, can be used to treat patients with acute or subacute Budd-Chiari syndrome who are symptomatic, provided a venous obstruction amenable to percutaneous angioplasty and stenting is visualized radiologically (eg, on magnetic resonance venography or percutaneous

venography). These interventional procedures, along with TIPS placement, are assuming a major role in the management of patients with Budd-Chiari syndrome [31-33]. (See "Budd-Chiari syndrome: Epidemiology, clinical manifestations, and diagnosis", section on 'Radiographic findings'.)

The anatomy of the hepatic venous circulation in some patients with Budd-Chiari syndrome may reveal a focal abnormality that is amenable to balloon angioplasty. A membranous web in the inferior vena cava, for example, can be dilated [26,34]. Balloon dilation of hepatic veins may also be effective in some patients [35]. Angioplasty can be combined with thrombolytic therapy in patients in whom there is recent thrombosis of a single hepatic vein [36].

The major problem with angioplasty for Budd-Chiari syndrome is that reocclusion of the affected vessel is common. Placement of an expandable metal stent in the occluded vessel following angioplasty may help to maintain patency [33,37-40]. Experience with this technique is limited to small numbers of patients; long-term survival with improvement in liver function and lessening of portal hypertension has been reported [38]. However, many patients eventually require liver transplantation. (See 'Liver transplantation' below.)

Once inserted, the stent cannot be removed. Therefore, placement should be coordinated with a liver transplantation team in patients who would otherwise be eligible for liver transplantation. Placement of a stent above the intrahepatic inferior vena cava, for example, complicates, and in some cases precludes, anastomosis of the donor and host inferior vena cava required during liver transplantation.

Decompress the liver — TIPS placement may be an option in patients with acute or subacute Budd-Chiari syndrome who are symptomatic and do not experience improvement with other treatments. TIPS may have a role as a temporizing measure to treat complications of portal hypertension (eg, variceal bleeding) prior to liver transplantation in patients with acute liver failure [41,42]. Finally, TIPS placement should be considered for patients with chronic Budd-Chiari syndrome and complications of portal hypertension. Alternative treatments if TIPS placement is not feasible or is unsuccessful include surgical shunts and liver transplantation.

However, TIPS placement may not be technically feasible because of the presence of thrombosis and because it is associated with high rates of shunt occlusion. Because of these limitations, among patients with acute or subacute Budd-Chiari syndrome, we reserve TIPS placement for patients for whom treatment with anticoagulation, thrombolysis, and/or angioplasty (with or without stenting) has failed. In particular, patients should be carefully evaluated to see if they are candidates for angioplasty and stenting prior to TIPS placement.

If TIPS is being used to treat complications of portal hypertension (eg, variceal bleeding) prior to liver transplantation in patients with acute liver failure, it is important that it be performed properly because failure to do so can result in complications such as portal vein thrombosis, which may greatly complicate or preclude subsequent liver transplantation.

Transjugular intrahepatic portosystemic shunt — The rationale behind TIPS placement in Budd-Chiari syndrome is to decompress congested segments in the liver by creating an alternative venous outflow tract. However, TIPS placement is not technically feasible in some patients, may only drain a small portion of the liver, and is associated with a high rate of occlusion [43]. The availability of polytetrafluoroethylene-covered stents, which have a reduced incidence of TIPS dysfunction, has led to increasing use of this intervention [9,44]. (See "Overview of transjugular intrahepatic portosystemic shunts (TIPS)".)

Several reports suggest that TIPS placement can be successfully performed in patients with Budd-Chiari syndrome [37,45-53]. In one of the largest series (which included patients with severe Budd-Chiari syndrome who did not respond to medical therapy or attempts at achieving recanalization), overall five-year survival after TIPS placement (84 percent) was similar to five-year survival from published reports of orthotopic liver transplantation for Budd-Chiari syndrome [51]. Transplant-free survival rates at one and five years after TIPS placement were 88 and 78 percent, respectively. A prognostic score based on age, bilirubin, and international normalized ratio levels predicted a small group of patients expected to have high mortality after TIPS, who would be better managed by liver transplantation.

Surgical therapy — The goal of surgical therapy for Budd-Chiari syndrome is to restore hepatic venous drainage and thereby decompress the liver. This is usually done by the creation of a surgical shunt. Surgical thrombectomy alone is usually not technically possible though it has been performed in conjunction with dorsocranial liver resection with good results in selected cases [54,55]. Surgical decompression is unlikely to be beneficial in patients who have cirrhosis or biochemical evidence of advanced liver dysfunction (elevated prothrombin time and serum bilirubin level, decreased serum albumin). Such patients are best managed with liver transplantation. (See 'Liver transplantation' below.)

Most surgical shunts drain the portal or mesenteric venous system into the inferior vena cava or another systemic vein. This allows blood entering the liver via the hepatic artery to have a low-pressure route by which to drain out of the liver. Severe acute hepatocellular necrosis caused by pressure-induced atrophy of hepatocytes will often regress following the creation of decompressive shunts.

Visceral arteriography is mandatory when planning surgery to decompress the congested liver. The hepatic arteries are usually stretched, arched, and attenuated, which may influence the surgical approach. In addition, the patency of the portal, splenic, and mesenteric veins can be determined, and tumors involving the liver, inferior vena cava, or the hepatic and portal veins can be identified.

Surgical shunts — Multiple surgical approaches have been attempted in the treatment of Budd-Chiari syndrome [56-61]. However, surgical shunt procedures are infrequently performed and have been largely replaced TIPS placement with a covered stent [16] (see 'Transjugular intrahepatic portosystemic shunt' above). For patients who undergo shunt surgery, multidisciplinary care should be coordinated with a liver transplantation team since liver transplantation may be required despite shunt surgery. The underlying cause of the thrombotic diathesis should be identified and treated prior to considering shunt surgery.

Side-to-side portacaval, splenorenal, and mesocaval shunts all are feasible only if the inferior vena cava is patent and without a significant pressure gradient between its infrahepatic and suprahepatic portions. It has been suggested that the infrahepatic pressure in the inferior vena cava should be at least 10 mmHg lower than the portal pressure [62]. A simple and direct side-to-side portacaval anastomosis may also be technically difficult in the presence of caudate lobe hypertrophy. Such patients often require a mesocaval shunt.

Synthetic shunts can be constructed from the portal-mesenteric system to the right atrium (meso-atrial shunt), which can bypass the inferior vena cava if it is occluded or there is a significant pressure gradient. However, shunts that require artificial graft material are more likely to be complicated by thrombosis. To overcome this complication, one group has tried the placement of a side-to-side portacaval shunt combined with a cavoatrial shunt through a Gore-Tex graft and has reported long-term patency [61].

Five-year survival following shunt surgery depends on the extent of liver damage prior to surgery and the continued patency of the shunt [61,63]. Five-year survival rates as high as 90 percent have been reported in patients who underwent shunt surgery prior to the development of cirrhosis, and whose shunts remained patent [5,61,63,64].

Perioperative management — Appropriate perioperative management of patients undergoing surgical decompression is critical [65]. Maintenance of shunt patency often requires anticoagulation. In addition, Doppler ultrasound should be performed periodically to evaluate shunt patency, particularly for shunts in which a synthetic graft has been used.

Clinical or biochemical deterioration in patients following shunt surgery should be investigated by angiography to determine whether the shunt has thrombosed, which may be corrected by

angioplasty.

Liver transplantation — Liver transplantation may be the only option for patients with Budd-Chiari syndrome who are not candidates for other treatments, who have decompensated cirrhosis, or for whom other treatments are ineffective. Patients who developed Budd-Chiari syndrome as a result of protein S, protein C, or antithrombin III deficiency may also be cured of their clotting tendency by liver transplantation since the transplanted liver produces normal amounts of these enzymes. (See "Liver transplantation in adults: Patient selection and pretransplantation evaluation".)

Survival following liver transplantation depends on the underlying cause of the Budd-Chiari syndrome and the patient's condition at the time of the transplantation [65-68]. Advances in liver transplantation technology and adoption of the Model for End-stage Liver Disease (MELD) score for deceased donor liver allocation may have led to improved survival following liver transplantation for Budd-Chiari syndrome [69] (see "Model for End-stage Liver Disease (MELD)"):

- One of the largest series (510 patients) compared survival in patients with Budd-Chiari syndrome who underwent liver transplantation after adoption of the MELD score with the pre-MELD era [69]. Three-year graft survival (81 versus 65 percent) and patient survival (85 versus 73 percent) were better following adoption of the MELD score.

- In another large series (248 patients), overall survival was 76, 71, and 68 percent at one, five, and 10 years, respectively [67]. Most deaths (77 percent) occurred in the first three months. The only predictors of mortality were impaired renal function and a history of a shunt.

- Ten-year survival ranged from 69 to 84 percent in two other series of patients undergoing liver transplantation [65,70]. In one report, vascular complications were more common in patients transplanted for Budd-Chiari syndrome than other indications [70]. Although survival was similar to or better than that of patients transplanted for other indications, these patients may have had less severe liver dysfunction prior to transplantation.

Manage complications — Patients with acute or chronic Budd-Chiari syndrome may develop portal hypertension. Manifestations of portal hypertension include ascites and variceal bleeding. Many patients will require a dietary sodium restriction, diuretics, and possibly paracentesis to control ascites. The management of the complications of portal hypertension is discussed elsewhere. (See "Cirrhosis in adults: Overview of complications, general management, and prognosis", section on 'Complications of portal hypertension'.)

Patients should also have a dietary history obtained, especially if there has been weight loss or other evidence of malnutrition. Nutritional therapy should be initiated if needed. (See "Evaluating nutritional status in adults with cirrhosis".)

FOLLOW-UP AND MONITORING

Patients in whom patency of the thrombosed vessel could not be achieved should be monitored closely for disease progression (ie, deterioration of hepatic synthetic function). In addition to laboratory studies (serum aminotransferases, bilirubin, albumin, international normalized ratio, complete blood count) every three months, a reasonable approach is to perform serial upper endoscopies (screening for varices) and liver biopsies annually, decreasing the interval to every two to three years if laboratory, endoscopic, and histologic findings are stable. It may be possible to decrease the frequency of liver biopsies by using noninvasive methods for detecting hepatic fibrosis (eg, ultrasound-based transient elastography) to evaluate for fibrosis or cirrhosis. Patients receiving long-term anticoagulation need close follow-up and monitoring of coagulation status, especially during invasive procedures [71]. (See "Noninvasive assessment of hepatic fibrosis: Overview of serologic tests and imaging examinations".)

Patients with compensated cirrhosis due to Budd-Chiari syndrome should be monitored for the development of complications from portal hypertension [72]. In addition, patients with cirrhosis should be monitored for the late development of hepatocellular carcinoma [72]. Finally, patients should be monitored for transformation of an underlying myeloproliferative neoplasm. (See "Surveillance for hepatocellular carcinoma in adults", section on 'Our approach to surveillance' and "Overview of the myeloproliferative neoplasms", section on 'Malignancies and disease transformation'.)

PREGNANCY

Budd-Chiari syndrome in women who are pregnant is discussed separately. (See "Overview of coincident acute hepatobiliary disease in pregnant women", section on 'Vascular disease'.)

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

- **Background** – Budd-Chiari syndrome is defined as hepatic venous outflow tract obstruction, independent of the level or mechanism of obstruction, provided the obstruction is not due to cardiac disease, pericardial disease, or sinusoidal obstruction syndrome (veno-occlusive disease). (See 'Introduction' above.)

- **Goals of therapy** – The approach to management depends on the patient's clinical and anatomic features, and local expertise. Treatment aims to (see 'Goals of therapy' above):

  - Prevent the propagation of the clot
  - Restore patency of thrombosed veins
  - Decompress the congested liver
  - Prevent or manage complications

- **Prognosis** – Without treatment, the prognosis is poor (survival rates of approximately 10 percent at three years). However, with treatment, five-year survival rates of approximately 75 percent have been reported. (See 'Natural history and prognosis' above.)

- **Management** – We use a stepwise approach for the management of patients with Budd-Chiari syndrome (algorithm 1) (see 'Management overview' above):

  - Underlying disorders that predisposed to the development of Budd-Chiari syndrome should be corrected if possible. (See 'Treat predisposing conditions' above.)

  - Complications of portal hypertension (eg, ascites, variceal bleeding) should be treated, if present. (See 'Manage complications' above.)

  - We recommend that nearly all patients be started on anticoagulation at the time of diagnosis (**Grade 1B**). However, the risk of anticoagulation should be considered, especially in patients who present with bleeding complications or those who have varices. Prior to initiating anticoagulation, we perform an upper endoscopy to screen for varices. We prefer to initially treat with low molecular weight heparin. We also start patients on an oral vitamin K antagonist (eg, warfarin). Once the international normalized ratio is between 2 and 3, we discontinue the low molecular weight heparin. Anticoagulation is continued indefinitely unless a major contraindication is present, a complication occurs, or the obstruction is due to an anatomic cause that has been corrected. (See 'Prevent propagation of the clot' above.)

- For patients with acute Budd-Chiari syndrome who have a well-defined clot that is known to be less than three to four weeks old, we suggest thrombolytic therapy rather than anticoagulation monotherapy (**Grade 2C**). We avoid using thrombolytic agents in patients who have an extensive clot involving the intrahepatic vena cava and hepatic veins, or in patients who have a clot of unknown age. (See 'Thrombolytic therapy' above.)

- For patients with acute or subacute Budd-Chiari syndrome who are symptomatic and are not candidates for thrombolytic therapy, we suggest angioplasty and stenting with anticoagulation rather than anticoagulation monotherapy, provided a venous obstruction amenable to percutaneous angioplasty and stenting is visualized radiologically (**Grade 2C**). (See 'Angioplasty and stenting' above.)

- Treatment options for patients with acute or subacute Budd-Chiari syndrome who fail to improve with other treatments include transjugular intrahepatic portosystemic shunt (TIPS) placement, surgical shunting, and liver transplantation. TIPS placement and liver transplantation may also be options for patients with cirrhosis who develop complications of cirrhosis. (See 'Decompress the liver' above.)

The management of patients with acute liver failure is discussed in detail elsewhere. (See "Acute liver failure in children: Management, complications, and outcomes".)

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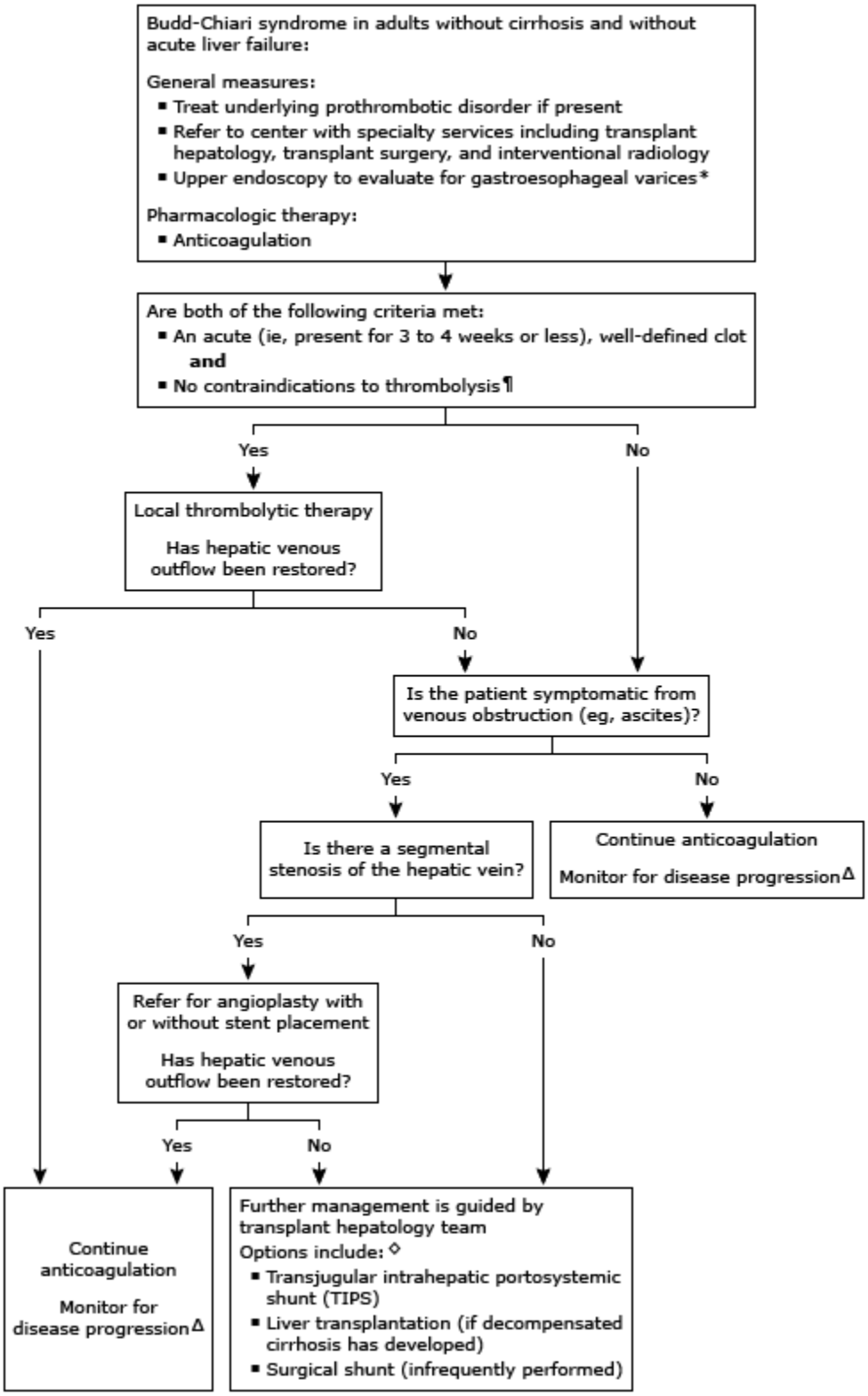
GRAPHICS

An approach to initial management of Budd-Chiari syndrome



Refer to UpToDate content on management of adults with Budd-Chiari syndrome for additional details.

HCC: hepatocellular carcinoma; TIPS: transjugular intrahepatic portosystemic shunt.

* For patients with gastroesophageal varices who are at risk for bleeding, we implement measures to prevent bleeding prior to starting anticoagulation. We typically use a nonselective beta blocker for prophylaxis. Refer to UpToDate content on prevention of variceal bleeding for additional details.

¶ Contraindications to thrombolytic therapy include large gastroesophageal varices and/or gastrointestinal bleeding.

Δ Disease progression includes the development of cirrhosis and its complications (eg, portal hypertension, HCC) and progression of an underlying myeloproliferative disorder.

◇ The choice of intervention is informed by several factors including technical feasibility for TIPS, candidacy for liver transplantation, underlying prothrombotic disorder, and local expertise. When feasible, TIPS is more commonly performed than surgical shunt because it is less invasive and is effective for patients who have not responded to other therapies. Liver transplantation may be an option for patients who develop decompensated cirrhosis and have not responded to endovascular intervention and anticoagulation.

Graphic 95281 Version 4.0

Prothrombotic risk factors for BCS

A. Acquired thrombophilia
▪ Myeloproliferative disease
▪ Polycythemia vera
▪ Essential thrombocytosis
▪ Idiopathic myelofibrosis
▪ *JAK2* V617F mutation
▪ Paroxysmal nocturnal hemoglobinuria
▪ Behçet disease
▪ Hyperhomocysteinemia
▪ Antiphospholipid syndrome
B. Inherited thrombophilia
▪ Factor V Leiden
▪ Prothrombin gene G20210A mutation
▪ MTHFR C677T mutation
▪ Thalassemia
▪ PC deficiency
▪ Protein S deficiency
▪ Antithrombin deficiency
C. Systemic factors
▪ Sarcoidosis
▪ Vasculitis
▪ Behçet disease
▪ Connective tissue disease
▪ Inflammatory bowel disease
D. Hormonal factors

▪ Recent oral contraceptive use
▪ Pregnancy

BCS: Budd-Chiari syndrome; *JAK2*: janus kinase 2; MTHFR: methyltetrahydrofolate; PC: protein C.

Graphic 130488 Version 1.0

Contributor Disclosures

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