# Budd-Chiari syndrome: Epidemiology, clinical manifestations, and diagnosis

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## INTRODUCTION

Budd-Chiari syndrome (BCS) 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 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 epidemiology, clinical manifestations, and diagnosis of Budd-Chiari syndrome. The etiology and treatment of Budd-Chiari syndrome are discussed separately. (See "Etiology of the Budd-Chiari syndrome" and "Budd-Chiari syndrome: Management".)

## EPIDEMIOLOGY

Studies suggest that in non-Asian countries, Budd-Chiari syndrome is more common in women and usually presents in the third or fourth decade of life (although it may occur in children or older adults) [1,2]. By contrast, in Asia, there is a slight predominance of men, with a median age of 45 years at presentation. In the non-Asian countries, pure hepatic vein blockage is more

common, whereas in Asia, pure inferior vena cava or combined inferior vena cava and hepatic vein blockage predominate [2,3].

In a ten year study of hospital admissions in Italy that included 287 patients with BCS, the incidence rates for males and females were 2.0 and 2.2 per million inhabitants, respectively [4]. The median age was 50 years (interquartile range 36 to 68 years) and 54 percent of patients were female. In another series that included 237 patients who had been treated for BCS at four centers in the United States, the Netherlands, and France, the median age was 35 years (range 13 to 76 years) and 67 percent of patients were female [3]. The location of the outflow obstruction was in the hepatic veins (62 percent), inferior vena cava (7 percent), or both (31 percent), and 34 patients (14 percent) had associated portal vein thrombosis.

## CATEGORIZATION

Budd-Chiari syndrome is categorized by disease duration and severity [5-7]:

- Acute (fulminant) Budd-Chiari syndrome with acute liver failure – Characterized by acute liver injury with elevated transaminases, jaundice, hepatic encephalopathy, and an elevated prothrombin time/international normalized ratio; hepatic encephalopathy develops within eight weeks after the development of jaundice.
- Acute Budd-Chiari syndrome but without liver failure – 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 (non-fulminant) liver disease have not yet developed venous collaterals, whereas venous collaterals are seen in patients with subacute and chronic liver disease.

## CLINICAL MANIFESTATIONS

Symptoms in patients with acute Budd-Chiari syndrome may often include abdominal pain, abdominal distension from ascites, hepatomegaly, and gastrointestinal bleeding (from varices or portal hypertensive gastropathy). Less common presenting symptoms include lower

extremity edema, jaundice, fever and/or hepatic encephalopathy [8]. Patients with subacute or chronic Budd-Chiari syndrome may be asymptomatic. In such patients, the hepatic venous outflow obstruction is often discovered as part of the evaluation of abnormal liver blood tests or when imaging is obtained for other reasons. (See 'Categorization' above.)

Acute liver failure is seen in approximately 5 percent of patients, whereas approximately 20 percent present with acute Budd-Chiari syndrome without acute liver failure [6,8-10]. While studies often group patients with subacute and chronic Budd-Chiari syndrome together, it is thought that the subacute presentation accounts for the majority of patients [5].

Because the presentation of Budd-Chiari syndrome is highly variable, clinicians should consider it in the differential diagnosis of patients presenting with acute liver failure, acute hepatitis, or chronic liver disease. (See 'When to consider Budd-Chiari syndrome' below.)

**Acute liver failure** — Acute liver failure is characterized by acute liver injury with elevated transaminases, jaundice, hepatic encephalopathy, and an elevated prothrombin time/international normalized ratio; hepatic encephalopathy develops within eight weeks after the development of jaundice. The clinical manifestations of acute liver failure are discussed in detail elsewhere. (See "Acute liver failure in adults: Etiology, clinical manifestations, and diagnosis", section on 'Clinical manifestations'.)

**Acute Budd-Chiari syndrome without liver failure** — In patients with acute Budd-Chiari syndrome, clinical manifestations develop rapidly (over the course of weeks). Patients usually present with severe right upper quadrant pain and hepatomegaly [3]. Jaundice and ascites may not be apparent initially but often develop rapidly. Variceal bleeding may also occur.

Among patients with an acute presentation, liver blood tests are abnormal although the magnitude and pattern of abnormalities can vary [5]. Elevation of serum aminotransferases occurs because vascular congestion results in ischemic hepatocellular damage. Serum aminotransferase concentrations can range from 100 to 200 int. units/L to more than 600 int. units/L. The serum alkaline phosphatase in acute Budd-Chiari syndrome is often in the range of 300 to 400 int. units/L and serum bilirubin levels are usually less than 7 mg/dL at the time of presentation, though they may subsequently increase.

Ascitic fluid in patients with Budd-Chiari syndrome has a high serum-to-ascites protein gradient (>1.1), reflecting elevated portal pressures. The protein concentration is variable (ranging from 1.5 to 4.9 g/dL in one report) and is directly related to the serum protein and inversely related to portal pressure [11]. (See "Evaluation of adults with ascites".)

**Subacute and chronic Budd-Chiari syndrome** — Patients with subacute or chronic Budd-Chiari syndrome may be asymptomatic or minimally symptomatic until the disease progresses [10]. It is estimated that 15 to 20 percent of patients are asymptomatic [2]. Patients who are asymptomatic often have large hepatic vein collaterals [10].

In subacute Budd-Chiari syndrome, symptoms develop over the course of months. Patients may report a history of vague discomfort in the mid epigastrium or right upper quadrant. Ascites and hepatic necrosis may be minimal due to decompression of the sinusoids by portal and hepatic venous collaterals. However, ascites and lower extremity edema may occur because chronic occlusion of the hepatic veins may be associated with hypertrophy of the caudate lobe of the liver, which receives a separate blood supply from the rest of the liver. The hypertrophied caudate lobe can eventually cause compression of the intrahepatic portion of the inferior vena cava, leading to further outflow obstruction and portal hypertension. Venous collaterals may be seen on the anterior abdominal wall.

In chronic Budd-Chiari syndrome, symptoms develop once the patient has cirrhosis. Patients who develop cirrhosis may have stigmata of chronic liver disease such as spider angiomata and palmar erythema. They may also have signs of portal hypertension such as ascites (which may be massive) and esophageal varices. Hepatomegaly and abdominal pain are also common, but encephalopathy is not. Hepatopulmonary syndrome has been described in up to 28 percent of patients [12]. (See "Cirrhosis in adults: Etiologies, clinical manifestations, and diagnosis", section on 'Clinical manifestations' and "Hepatopulmonary syndrome in adults: Prevalence, causes, clinical manifestations, and diagnosis".)

Patients with subacute or chronic Budd-Chiari syndrome may present with normal or mild to moderate elevations of serum aminotransferases, alkaline phosphatase, and serum bilirubin [5]. However, hyperbilirubinemia resulting in jaundice is rare. They may also have hypoalbuminemia. As in patients with acute Budd-Chiari syndrome, ascitic fluid in patients with subacute or chronic Budd-Chiari syndrome has a high serum-to-ascites protein gradient (>1.1).

## DIAGNOSIS

A diagnosis of Budd-Chiari syndrome can usually be established with Doppler ultrasonography. Patients should also undergo an evaluation for conditions that may have predisposed to the development of Budd-Chiari syndrome such as prothrombotic disorders. (See 'Evaluating for predisposing conditions' below.)

**When to consider Budd-Chiari syndrome** — Because the presentation of Budd-Chiari syndrome is highly variable, clinicians should consider it in the differential diagnosis of patients presenting with acute liver failure, acute hepatitis, or chronic liver disease, particularly if the patient has known risk factors for Budd-Chiari syndrome ( table 1). Among patients with acute liver failure, the presence of hepatomegaly, right upper quadrant pain, and ascites should increase the suspicion for Budd-Chiari syndrome. (See "Etiology of the Budd-Chiari syndrome".)

### Establishing the diagnosis

**General approach** — The diagnosis of Budd-Chiari syndrome can usually be established noninvasively with Doppler ultrasonography. The portal and splenic circulation should also be evaluated to exclude concurrent portal or splenic vein thrombosis.

Computed tomography (CT) or magnetic resonance imaging (MRI) can be performed to confirm the diagnosis, to aid with treatment planning, or if an experienced Doppler sonographer is not available. In addition, CT or MRI can be performed in patients with an unremarkable ultrasound examination but in whom the suspicion for Budd-Chiari syndrome is high (eg, a patient with a thrombophilic disorder and acute hepatitis whose evaluation for other causes of hepatitis is negative). Venography should be performed if noninvasive tests are negative or nondiagnostic, but there is still strong clinical suspicion for the disease. In addition, venography can be used to confirm the diagnosis and to plan therapeutic interventions. (See "Budd-Chiari syndrome: Management", section on 'Management'.)

Doppler ultrasonography is also used to confirm the diagnosis if a patient's initial imaging study is a CT scan or MRI that suggests Budd-Chiari syndrome.

Liver biopsy is generally not required to diagnose Budd-Chiari syndrome. In addition, it should be kept in mind that patients are at risk of bleeding from the biopsy site, particularly if started on anticoagulation, which is a primary element of the treatment for Budd-Chiari syndrome. (See "Budd-Chiari syndrome: Management", section on 'Angioplasty and stenting'.)

#### Radiographic findings

**Doppler ultrasonography** — Findings on Doppler ultrasonography in patients with Budd-Chiari syndrome correlate well with pathologic and venographic findings [13-16]. In addition, Doppler ultrasonography is widely available and does not require contrast administration or exposure to radiation. Nonspecific ultrasonographic findings include hepatomegaly, splenomegaly, ascites, intra-abdominal collaterals, caudate lobe hypertrophy, atrophy of other hepatic lobes, and compression or narrowing of the inferior vena cava.

Findings that are more specific for hepatic venous outflow obstruction include [2,17]:

- Inability to visualize the junction of the major hepatic veins with the inferior vena cava
- Thickening, irregularity, stenosis, or dilation of the walls of the hepatic veins
- A spider-web appearance in the vicinity of the hepatic vein ostia coupled with the absence of a normal hepatic vein
- A hyperechoic cord replacing a normal vein
- Abnormal flow patterns seen on Doppler imaging, including:
  - A large hepatic vein that appears void of flow-signal or has reversed or turbulent flow
  - Large intrahepatic or subcapsular collaterals with continuous flow connecting the hepatic veins or the diaphragmatic or intercostal veins
  - A hepatic wave form that is flat or lacks fluttering

**CT scan or MRI** — Contrast-enhanced CT scan may reveal the same nonspecific abnormalities seen during ultrasonography ( image 1). More specific findings on CT scan that suggest Budd-Chiari syndrome include:

- Delayed or absent filling of the three major hepatic veins (which are usually visible within 40 to 60 seconds after rapid intravenous contrast).
- A patchy, flea-bitten appearance of the liver due to increased central contrast enhancement relative to the periphery.
- Rapid clearance of contrast from the caudate lobe.
- Narrowing and/or lack of opacification of the inferior vena cava.

Contrast enhanced-MRI is also useful in the diagnosis of Budd-Chiari syndrome [18,19]. In addition to the nonspecific findings described above, the absence or reduction in caliber of hepatic veins and the typical distorted "comma-shaped" intrahepatic collaterals are easily demonstrable.

**Venography** — Venography should be performed if noninvasive tests are negative or nondiagnostic, but there is strong clinical suspicion for the disease. It can also be used to direct subsequent therapy by clearly defining which vessels are involved. The gold standard for diagnosing Budd-Chiari syndrome is hepatic venography, which is performed by accessing the

hepatic venous circulation percutaneously via the internal jugular vein, cephalic vein, or femoral vein.

The examination should include venous pressure measurements above and below the entrance of the hepatic veins into the inferior vena cava to determine whether a significant pressure gradient exists. Once the inferior vena cava has been shown to be patent, opacification of each of the individual hepatic veins should be attempted. However, venography of some or all of the hepatic veins may not be feasible in many patients with Budd-Chiari syndrome. Injection of contrast following balloon occlusion of a specific hepatic vein may facilitate its visualization and also demonstrate the so-called "spider-web" pattern characteristic of the Budd-Chiari syndrome. This pattern depicts the attempt of the liver to form venous collaterals to bypass the occluded hepatic veins.

Venography can be critical for directing therapy. Noninvasive studies may not accurately define the extent or characteristics of the hepatic venous flow. In particular, compression or occlusion of the intrahepatic vena cava leads to sluggish flow in hepatic veins. As a result, hepatic veins that are patent and amenable to therapy may be undetectable on Doppler imaging. MRI venography can be helpful in better defining the venous anatomy.

**Liver biopsy** — Liver biopsy is rarely required for the diagnosis of Budd-Chiari syndrome. We generally perform a liver biopsy when:

- There is confusion regarding the diagnosis (an uncommon situation given the multitude of available imaging tests).
- In selected patients with a subacute presentation, when the presence of cirrhosis is not apparent by noninvasive studies, and the finding of significant fibrosis/cirrhosis or severe centrizonal congestion would indicate that the patient could benefit from transjugular intrahepatic or surgical portosystemic shunting.

While a liver biopsy may diagnose Budd-Chiari syndrome, particularly in the acute or subacute form of the disease, the diagnosis can usually be made noninvasively. Histologic features of Budd-Chiari syndrome include centrizonal congestion, necrosis, and hemorrhage. Large regenerative nodules, obstructive portal venopathy, and fibrosis/cirrhosis may also be found [20].

The thrombotic process in Budd-Chiari syndrome may not involve all the hepatic veins. Thus, the distribution of the typical pathologic findings may be focal or patchy. As a result, some patients require biopsy of both the right and the left lobes of the liver. A laparoscopic approach may be better suited for this purpose.

It is not clear if histologic findings at the time of diagnosis help predict survival. In one study, histologic findings did not predict five-year survival, though there was a trend toward decreased survival among those with central vein thrombosis [21].

Liver biopsies are often obtained via the transjugular route during performance of hepatic venography since the majority of patients have ascites and will require anticoagulation, which increases the risk of the percutaneous approach. (See "Transjugular liver biopsy", section on 'Indications and contraindications'.)

**Evaluating for predisposing conditions** — An underlying disorder can be identified in over 80 percent of patients with Budd-Chiari syndrome [8,22-24]. Many of these disorders are characterized by a hypercoagulable state ( table 1). More than one thrombotic risk factor is present in a quarter of patients [5]. (See "Etiology of the Budd-Chiari syndrome".)

The work up includes [25,26]:

- Ultrasonography, CT scan, or MRI to look for space-occupying lesions or malignant tumors compressing or invading the hepatic venous outflow tract.
- Evaluation for acquired and inherited thrombotic conditions (see "Evaluating adult patients with established venous thromboembolism for acquired and inherited risk factors").
- Evaluation for systemic diseases associated with Budd-Chiari syndrome such as ulcerative colitis and celiac disease.

## DIFFERENTIAL DIAGNOSIS

Budd-Chiari syndrome shares features with other causes of acute or chronic liver disease including viral hepatitis, alcohol-associated liver disease, nonalcoholic fatty liver disease, hemochromatosis, ischemic hepatitis, right-sided heart failure, and drug-induced liver injury. Because these disorders have similar presentations, testing should be undertaken to rule out other common causes of acute (including acute liver failure), subacute, or chronic liver disease. Of note, signs of right-sided congestive heart failure (such as jugular venous distension) are not characteristic of Budd-Chiari syndrome and suggest an underlying cardiac cause of fluid accumulation.

The approach to the evaluation of patients with acute or chronic liver disease is discussed in detail elsewhere. (See "Approach to the patient with abnormal liver biochemical and function tests", section on 'Initial evaluation' and "Acute liver failure in adults: Etiology, clinical

manifestations, and diagnosis", section on 'Determining the cause of acute liver failure' and "Heart failure: Clinical manifestations and diagnosis in adults".)

## SPECIAL POPULATIONS

**Pediatrics** — Budd-Chiari syndrome is rare in children. In a retrospective study including pediatric patients with liver disease who were evaluated during a 15-year period, the prevalence of BCS was less than 0.1 percent (seven patients) [27]. Risk factors for thrombosis were present in all children who ranged in age from 2 to 17 years at the time of diagnosis. Risk factors for venous thrombosis in children, including inherited thrombophilia, are discussed separately. (See "Venous thrombosis and thromboembolism (VTE) in children: Risk factors, clinical manifestations, and diagnosis", section on 'Risk factors'.)

**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). Primary Budd-Chiari syndrome is present when there is obstruction due to a primarily venous process (thrombosis or phlebitis), whereas secondary Budd-Chiari 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. (See 'Introduction' above.)

- **Patient presentation** – Patients with Budd-Chiari syndrome may present with evidence of acute liver failure. More often, they present with acute liver disease (rapid symptom onset without liver failure), subacute liver disease (insidious symptom onset prior to the

development of cirrhosis), or chronic liver disease (symptom onset once cirrhosis has developed). Patients with acute liver failure or acute (non-fulminant) liver disease have not yet developed venous collaterals, whereas venous collaterals are seen in patients with subacute and chronic liver disease. (See 'Categorization' above.)

- **Clinical manifestations** – Symptoms in patients with acute Budd-Chiari syndrome may often include abdominal pain, abdominal distension from ascites, hepatomegaly, and gastrointestinal bleeding (from varices or portal hypertensive gastropathy). Less common presenting symptoms include lower extremity edema, jaundice, fever, and/or hepatic encephalopathy. Patients with subacute or chronic Budd-Chiari syndrome may be asymptomatic. In such patients, the hepatic venous outflow obstruction is often discovered as part of the evaluation of abnormal liver blood tests or when imaging is obtained for other reasons. (See 'Categorization' above and 'Clinical manifestations' above.)

- **Diagnosis** – Because the presentation of Budd-Chiari syndrome is highly variable, clinicians should consider it in the differential diagnosis of patients presenting with acute liver failure, acute hepatitis, or chronic liver disease, particularly if the patient has known risk factors for Budd-Chiari syndrome ( table 1). (See 'When to consider Budd-Chiari syndrome' above.)

  The diagnosis of Budd-Chiari syndrome can usually be established noninvasively with Doppler ultrasonography. The portal and splenic circulation should also be evaluated to exclude concurrent thrombosis. (See 'Diagnosis' above.)

  Contrast-enhanced computed tomography (CT) or magnetic resonance imaging (MRI) can be performed to confirm the diagnosis, to aid with treatment planning, or if an experienced Doppler sonographer is not available. In addition, CT or MRI can be performed in patients with an unremarkable ultrasound examination but in whom the suspicion for Budd-Chiari syndrome is high. Venography should be performed if noninvasive tests are negative or nondiagnostic, but there is strong clinical suspicion for the disease. In addition, venography can be used to confirm the diagnosis and to plan therapeutic interventions. Liver biopsy is generally not required to diagnose Budd-Chiari syndrome. (See 'General approach' above.)

- **Evaluating for predisposing conditions** – An underlying disorder can be identified in over 80 percent of patients with the Budd-Chiari syndrome, so once Budd-Chiari syndrome is diagnosed, patients should undergo an evaluation for predisposing conditions. (See 'Evaluating for predisposing conditions' above.)

## ACKNOWLEDGMENT

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### REFERENCES

1. Mahmoud AE, Mendoza A, Meshikhes AN, et al. Clinical spectrum, investigations and treatment of Budd-Chiari syndrome. QJM 1996; 89:37.
2. Plessier A, Valla DC. Budd-Chiari syndrome. Semin Liver Dis 2008; 28:259.
3. Darwish Murad S, Valla DC, de Groen PC, et al. Determinants of survival and the effect of portosystemic shunting in patients with Budd-Chiari syndrome. Hepatology 2004; 39:500.
4. Ageno W, Dentali F, Pomero F, et al. Incidence rates and case fatality rates of portal vein thrombosis and Budd-Chiari Syndrome. Thromb Haemost 2017; 117:794.
5. Menon KV, Shah V, Kamath PS. The Budd-Chiari syndrome. N Engl J Med 2004; 350:578.
6. Ferral H, Behrens G, Lopera J. Budd-Chiari syndrome. AJR Am J Roentgenol 2012; 199:737.
7. Gavriilidis P, Marangoni G, Ahmad J, Azoulay D. State of the Art, Current Perspectives, and Controversies of Budd-Chiari Syndrome: A Review. J Clin Med Res 2022; 14:147.
8. Darwish Murad S, Plessier A, Hernandez-Guerra M, et al. Etiology, management, and outcome of the Budd-Chiari syndrome. Ann Intern Med 2009; 151:167.
9. Fickert P, Ramschak H, Kenner L, et al. Acute Budd-Chiari syndrome with fulminant hepatic failure in a pregnant woman with factor V Leiden mutation. Gastroenterology 1996; 111:1670.
10. Hadengue A, Poliquin M, Vilgrain V, et al. The changing scene of hepatic vein thrombosis: recognition of asymptomatic cases. Gastroenterology 1994; 106:1042.
11. Mitchell MC, Boitnott JK, Kaufman S, et al. Budd-Chiari syndrome: etiology, diagnosis and management. Medicine (Baltimore) 1982; 61:199.
12. De BK, Sen S, Biswas PK, et al. Occurrence of hepatopulmonary syndrome in Budd-Chiari syndrome and the role of venous decompression. Gastroenterology 2002; 122:897.
13. Miller WJ, Federle MP, Straub WH, Davis PL. Budd-Chiari syndrome: imaging with pathologic correlation. Abdom Imaging 1993; 18:329.

14. Millener P, Grant EG, Rose S, et al. Color Doppler imaging findings in patients with Budd-Chiari syndrome: correlation with venographic findings. AJR Am J Roentgenol 1993; 161:307.
15. Chawla Y, Kumar S, Dhiman RK, et al. Duplex Doppler sonography in patients with Budd-Chiari syndrome. J Gastroenterol Hepatol 1999; 14:904.
16. Grant EG, Perrella R, Tessler FN, et al. Budd-Chiari syndrome: the results of duplex and color Doppler imaging. AJR Am J Roentgenol 1989; 152:377.
17. Faraoun SA, Boudjella Mel A, Debzi N, et al. Budd-Chiari syndrome: a prospective analysis of hepatic vein obstruction on ultrasonography, multidetector-row computed tomography and MR imaging. Abdom Imaging 2015; 40:1500.
18. Soyer P, Rabenandrasana A, Barge J, et al. MRI of Budd-Chiari syndrome. Abdom Imaging 1994; 19:325.
19. Friedman AC, Ramchandani P, Black M, et al. Magnetic resonance imaging diagnosis of Budd-Chiari syndrome. Gastroenterology 1986; 91:1289.
20. 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; 37:510.
21. 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; 35:338.
22. 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; 73:21.
23. Case records of the Massachusetts General Hospital. Weekly clinicopathological exercises. Case 51-1987. Progressive abdominal distention in a 51-year-old woman with polycythemia vera. N Engl J Med 1987; 317:1587.
24. Garcia-Pagán JC, Valla DC. Primary Budd-Chiari Syndrome. N Engl J Med 2023; 388:1307.
25. Simonetto DA, Singal AK, Garcia-Tsao G, et al. ACG Clinical Guideline: Disorders of the Hepatic and Mesenteric Circulation. Am J Gastroenterol 2020; 115:18.
26. Northup PG, Garcia-Pagan JC, Garcia-Tsao G, et al. Vascular Liver Disorders, Portal Vein Thrombosis, and Procedural Bleeding in Patients With Liver Disease: 2020 Practice Guidance by the American Association for the Study of Liver Diseases. Hepatology 2021; 73:366.
27. Nobre S, Khanna R, Bab N, et al. Primary Budd-Chiari Syndrome in Children: King's College Hospital Experience. J Pediatr Gastroenterol Nutr 2017; 65:93.

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## GRAPHICS

### Prothrombotic risk factors for BCS

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|---|
| **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 |

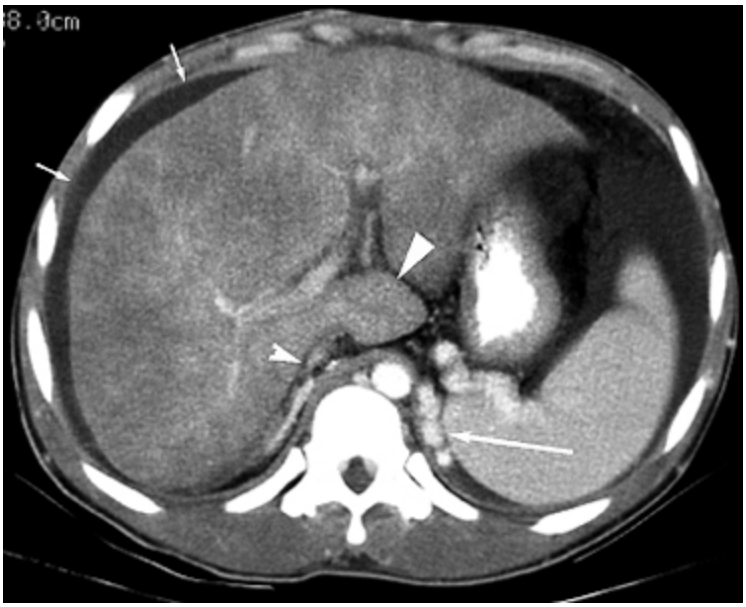
| **D. Hormonal factors** |
| --- |
| ▪ Recent oral contraceptive use |
| ▪ Pregnancy |

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



Graphic 130488 Version 1.0

## Budd-Chiari syndrome on computed tomography scan




Computed tomography (CT) scan through the liver of a 17-year-old male with new onset abdominal distension shows a mottled appearance to the underperfused liver with collapsed portal veins, ascites (small arrows), and extensive retroperitoneal varices (large arrow). Note also the enlarged caudate lobe of the liver (large arrowhead) and the collapsed small inferior vena cava (small arrowhead). These are all characteristic imaging features of the Budd-Chiari syndrome.

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*Courtesy of Jonathan Kruskal, MD.*

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Graphic 77769 Version 3.0

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