

(Table 8). The diagnosis requires demonstration of hepatic venous flow obstruction, and noninvasive imaging techniques (Doppler ultrasonography, CT, or MR) are the mainstay of diagnosis. Doppler ultrasound, performed by an experienced operator, has a sensitivity >75%.⁽²⁰⁹⁾ Typical ultrasound features of venous obstruction are identification of thrombus, nonvisualization of the HV, collateral veins and transformation of the HV into a cord lacking flow signals, caudate lobe hypertrophy, and a caudate vein >3 mm in diameter. Usually, the role of contrast-enhanced MR or CT is for diagnosis confirmation and is the diagnostic study of choice in the absence of an experienced ultrasound operator. CT and MR can additionally depict rapid clearance of contrast from the caudate lobe and patchy hepatic enhancement attributable to uneven portal perfusion. Hepatic venography may be helpful in cases of uncertain diagnosis, and the most typical sign is the presence of a “spider web” pattern of collateral circulation. Liver biopsy is not necessary for diagnosis unless there is concern for small intrahepatic vein obstruction when histology is the only way to achieve definitive diagnosis.

Treatment Options

The goals of HVT/BCS treatment include management of portal hypertension, treatment of the underlying thrombotic or malignant disease, and restoration of hepatic venous outflow. The most recommended and supported treatment approach is a progressive therapeutic strategy,^(201,210) stepping from less invasive (medical) to more invasive (LT) treatment according to the clinical response. A major challenge is recognizing the appropriate time to step up to the next treatment choice in a given patient. Because of this difficulty, these patients should routinely be managed in tertiary referral centers.

Medical Therapies

Medical treatment must be initiated early in the course and include treatment of the complications of portal hypertension. In addition to therapeutic anticoagulation, early recognition and treatment of an underlying prothrombotic disorder is also critical. All patients with HVT/BCS, even in the absence of a

recognized prothrombotic disorder, should receive anticoagulation with the aim of achieving recanalization and preventing thrombosis progression. Immediate LMWH, followed by a VKA, is the most frequent anticoagulation approach. Long-term UFH should be avoided because of the risk of heparin-induced thrombocytopenia.⁽²¹⁰⁾ Although promising and clinically attractive, data with DOACs are very limited.⁽¹⁷⁸⁾ One quarter of HVT/BCS patients will be alive at 5 years with medical therapy alone.⁽²⁰¹⁾

Interventional Vascular Procedures

THROMBOLYSIS

The experience of thrombolysis in HVT/BCS is limited. In selected cases of recent and incomplete thrombosis, local instillation of recombinant tPA, streptokinase, or urokinase, usually in combination with angioplasty/stenting, can restore venous outflow.⁽²¹¹⁾ Known contraindications to thrombolytic therapy, as outlined in product package inserts, should be thoroughly investigated before consideration of this therapy.

PERCUTANEOUS ANGIOPLASTY

In cases of segmental stenosis, either in the cranial part of the HV or suprahepatic IVC,^(212,213) percutaneous transluminal angioplasty with or without stenting may restore HV outflow. This presentation only accounts for ~10% of the cases in Western populations,⁽²⁰¹⁾ but segmental stenosis occurs in >80% of patients in China.^(212,214) A single randomized controlled trial suggested that angioplasty plus routine stenting should be the first-line invasive therapy to treat short-length stenosis because this treatment is associated with a much lower incidence of restenosis than angioplasty alone.⁽²¹⁵⁾ However, the lack of survival difference and a high risk of selection bias in the population included in the study reduce the generalizability of this proposal, and therefore vein stenting should be reserved for angioplasty failures.⁽²¹⁶⁾ In addition, retrievable stents (retrieved after a median of 15 days) have also been proposed as an alternate treatment. However, these results should be interpreted