REVIEW ARTICLE

Budd–Chiari syndrome: illustrated review of current management

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Keywords
Budd–Chiari – hepatic vein thrombosis – liver transplantation – portasystemic shunt, surgical – portasystemic shunt, transjugular intrahepatic – review

Abstract
Budd–Chiari syndrome (BCS) is characterized by hepatic venous outflow obstruction at any level from the small hepatic veins to the atrio caval junction. BCS is a complex disease with a wide spectrum of aetiologies and presentations. This article reviews the current literature with respect to presentation, management and prognosis of the disease. Medical, interventional and surgical management of BCS is discussed. Particular attention is paid to interventional and surgical aspects of management. The review is augmented by images, which provide a clinical corollary to the text.

In 1845, an internist named George Budd described the classic triad of abdominal pain, hepatomegaly and ascites. In 1899, pathologist Hans Chiari documented the histopathological features of what is now known as the Budd–Chiari syndrome (BCS) (1, 2). BCS is characterized by hepatic venous outflow obstruction at any level from the small hepatic veins to the atrio caval junction, regardless of the cause of obstruction (3, 4).

The natural history of BCS is poorly understood (5). Described treatment modalities for BCS include anticoagulation and treatment of underlying disease, percutaneous interventional techniques, transjugular intrahepatic portosystemic shunt (TIPS), surgical shunts and liver transplantation. Because of the rarity of BCS, there have been no randomized prospective trials comparing medical with surgical treatment of this disease (3). Currently, clinical judgment and local expertise play a large role in the management of BCS. This review article discusses the presentation, management and prognosis of BCS with particular attention focused on the interventional and surgical aspects of treatment.

Pathophysiology
Obstruction is usually caused by a thrombus, but may result from extrinsic compression (tumour, abscess, cysts), membranous webs within the inferior vena cava (IVC) (3), or postoperative complications following liver transplantation (6–8). Membranous web occlusion of the IVC is much more common in the Asian population than in Western countries (1, 9). Membranous web occlusion was once thought to be a congenital lesion, but more recent evidence suggests that membranous webs are the sequelae of thrombus formation (10–12). Additionally, membranous webs have been associated with hepatocellular carcinoma (11). Posthepatic obstruction leads to increased sinusoidal pressure, sinusoidal congestion, hepatomegaly, hepatic pain, portal hypertension and ascites. Without venous outflow, the elevated sinusoidal pressure leads to perisinusoidal necrosis of hepatocytes in Rappaport zone 3 and eventually to liver failure (3). Ischaemic injury is compounded by oxidative injury caused by the release of free radicals from sinusoidal cells (13). BCS results from the occlusion of three hepatic veins in about 2/3 of the cases, isolated occlusion of the IVC.
in 10% of cases and combined occlusion in almost 1/3 of the cases (1). Caudate lobe hypertrophy (Fig. 1), which may further compress the IVC, can occur in up to 50% of chronic presentations (1).

Presentation

The majority of patients with BCS present with hepatomegaly, right upper quadrant pain and abdominal ascites (5, 14). Lower extremity oedema is another common finding on physical exam. Some patients may feature signs of portal hypertension such as variceal bleeding and refractory ascites without a significant impact on liver function. A high serum–ascites gradient (≥1.1 g/dl) is usually found (1).

Young children (<10 years of age) account for 1–7% of all cases of BCS. This group often presents late in the course of disease, by which time irreversible pathology may be present (15).

The most common underlying disorders in patients with BCS are myeloproliferative disorders such as polycythaemia vera and essential thrombocytosis (1). The prevalence of myeloproliferative disorders in this population can be as high as 53%. In Western nations, factor V Leiden (25%) and factor II gene mutation (5%) are other common aetiologies. Some patients may have a combination of thrombophilias (5). Other common associations include oral contraceptive use, protein C and S deficiencies, antiphospholipid syndrome, antithrombin III deficiency, paroxysmal noc-
turnal haemoglobinuria, cancer, Bechet’s syndrome and trauma (16). BCS after pregnancy has been described and is associated with a particularly poor prognosis (17). Twenty per cent of cases of BCS are idiopathic (1).

The Budd–Chiari syndrome is termed primary or secondary based on the aetiology and site of venous outflow obstruction. Primary BCS originates from within the lumen of the veins or venules and results from thrombus, webs or endophlebitis. Secondary BCS results from an extra-luminal lesion such as tumour, abscess or cyst, which can invade the lumen or cause extrinsic compression of the hepatic venous outflow tract (4).

Patients with BCS present with varying degrees of symptomatology and acuity. The clinical presentation of patients with BCS is governed by the extent and swiftness of hepatic outflow obstruction juxtaposed to the body’s ability to decompress the liver via development of collateral blood flow. Based on this pathophysiology, patients can be divided into the following categories: fulminant, acute, subacute or chronic (13). The subacute form is the most common presentation and is characterized by slow accumulation of ascites. Hepatocellular damage is minimal due to the development of hepatic and portal venous collaterals. The fulminant form is rare and is defined as the development of hepatic encephalopathy within 8 weeks of the onset of jaundice. Acute BCS has a rapid onset, intractable ascites and a short duration of symptoms. Collateral venous channels do not develop in the acute form of BCS. Chronic BCS usually occurs in the background of cirrhosis. A report by Dilawari included 177 patients with BCS and used a variation of the above classification scheme. This patient population presented with the following distribution: 7% acute fulminant, 28% acute and 65% chronic (18).

Diagnostic evaluation

Work-up of suspected BCS begins with imaging modalities such as Doppler ultrasound, computed Fig. 1. Coronal computed tomography (CT) showing caudate lobe hypertrophy (white arrow) and inferior vena cava (IVC) compression (black arrow).
tomography (CT) scan and magnetic resonance imaging (MRI). Radiographic analysis should attempt to determine the patency of the hepatic veins, IVC and portal vein because this information will determine therapeutic options. Other radiographic findings seen in BCS include inhomogeneous parenchymal enhancement, presence of intrahepatic collaterals, hypervascular nodules and caudate lobe hypertrophy (Fig. 1) (20). Doppler ultrasound, often the first study obtained, has a sensitivity and specificity as high as 85% (21). CT scans allow for the evaluation of ascites, hepatic vein patency, vena cava patency and caudate lobe hypertrophy. MRI may differentiate chronic from acute disease and provide further delineation of vascular anatomy. On MRI, patients with chronic BCS demonstrate heterogeneous signal intensity throughout the liver, while those with acute BCS show divergent signal intensities and decreased contrast enhancement in the periphery relative to the central region of the liver (22). Other investigations that may aid in management include infra- and suprahepatic caval pressures (Fig. 2), hepatic venography and liver biopsy (16). Imaging studies also play an important role in postoperative evaluation and surveillance of surgical shunts (Fig. 3).

Underlying hypercoagulable states should be investigated in all patients (16). Patients found to have membranous obstruction should also undergo hypercoagulable work-up, as recent evidence suggests that thrombosis may be the inciting event for membranous web formation (10–12). Of note, low levels of antithrombin III, protein C and protein S may be seen in patients with any disease resulting in hepatocellular dysfunction. Bone marrow biopsy should be considered to identify occult myeloproliferative diseases (23). Some authors recommend testing for JAK2 mutations in all patients with BCS in order to uncover a possible latent myeloproliferative disorder (1).

Liver biopsy may reveal a wide range of histological findings from sinusoidal congestion and inflammation to cirrhosis and parenchymal necrosis. Extravasation
of red cells into the liver-cell plate and space of Disse is a characteristic feature of BCS. Sample error during biopsy may occur as parenchymal changes may be heterogeneous. No correlation has been observed between histological findings and patient outcomes (16, 19, 24). Moreover, some authors argue that biopsy results should not play a dominant role in decision making because patients with fibrosis at initial presentation may do well with procedures other than transplantation (25).

Management strategies

Treatment of BCS generally follows a least invasive to most invasive strategy. However, this paradigm is not absolute and circumstances may dictate immediate surgery or even transplantation depending on the presentation. Algorithms for management of BCS have been reported (26, 27).

Plessier’s algorithm, which was applied to 51 patients over a 7-year period, resulted in overall 1-, 3- and 5-year survival rates of 96%, 89% and 89% respectively (27). This algorithm included medical management, recanalization, TIPS and liver transplant. Surgical shunts were not part of the algorithm.

Medical management

Medical management consists of anticoagulation, sodium restriction, diuretic therapy and paracentesis. Anticoagulation can be achieved with heparin in the acute setting and with warfarin for long-term treatment. Sodium should be restricted to approximately 60–90 mEq/day (1500–2000 mg of salt). Moderate ascites can be treated with oral diuretics such as furosemide (20–40 mg/day) and spironolactone (50–200 mg/day). Patients with symptomatic large-volume ascites can undergo periodic abdominal paracentesis (16). Some clinicians recommend medical therapy alone for patients with few symptoms, relatively normal liver-function tests and easily controlled ascites (13).

A report by McCarthy in 1985 showed that medical management alone resulted in death within 6 months in 12 of 14 patients (28). However, Khuroo reported more promising results from a case series where 8 of 20 patients treated with medical therapy alone had significant clinical and biochemical improvement with 53% of all medically treated patients alive after 24 months (29). Both these studies were retrospective case series from a single institution.

Plessier reported their experience with 51 cases of BCS and found that nine of their patients were successfully treated with medical therapy alone. Interestingly, this study found a 15% rate of heparin-induced thrombocytopenia, which is 10-fold higher than the rates from large clinical trials (27).

For patients who present with BCS several months after liver transplantation, medical management may be a viable option. Parrilla’s series reported that only three out of 1112 transplants suffered delayed hepatic outflow obstruction, and all three patients were successfully managed with diuretics for control of their ascites (7).

Interventional techniques and transjugular intrahepatic portosystemic shunt

In recent years, percutaneous interventions have played an increasing role in the management of patients with BCS. Indeed, most institutions are more likely to possess the capability to perform TIPS than to employ a surgeon with the expertise to perform a mesenteric-systemic shunt (9). Consequently, TIPS has become the first option when a shunting procedure is indicated (30). Catheter-directed thrombolytic therapy, angioplasty and stent placement can be effective in the acute setting (16), while TIPS may be employed in the acute or chronic setting.

A recent 10 patient case series and literature review by Sharma concluded that systemic thrombolytic therapy is of little value and that catheter-directed thrombolysis is more efficacious for acute thrombus that is not completely occlusive. The authors also felt that tissue plasminogen activator is the preferred agent and that it should be delivered just proximal to or within the thrombus (31).

Angioplasty and stenting are additional adjuncts that may be used. Angioplasty often suffers from high reocclusion rates. Thus, placement of stents in the IVC or hepatic veins has been recommended (Fig. 4) (31). A study out of China recently reported a case series of 115 in which stents were placed in the IVC and hepatic veins with success rates of 94% and 87% respectively. Patency reached 90% after a mean follow-up of more than 45 months (32). The proportion of patients with membranous occlusion, which may be more amenable to angioplasty and stenting, was much more prevalent in this population (57%) than would be expected in most Western institutions.

Angioplasty and stenting may offer certain advantages to patients who suffer from BCS after liver transplantation. The aetiology in this subset of patients is usually secondary to technical problems (33). Assuming hepatic function has not been lost, correction of the technical issue may be accomplished by percutaneous means and may obviate the need for...
more invasive procedures (34). Clinical improvement and long-term durability have been reported in small case series (8).

Unfortunately, most patients do not present acutely and their disease is often not amenable to thrombolitics, angioplasty or stenting. For these patients who present weeks to months after formation of hepatic vein thrombus, TIPS has become an attractive option in the elective and emergent situations (16). TIPS effectively decompresses the portal system and may serve as a bridge to transplant. The first report of TIPS for the treatment of BCS was in 1993 (35), and since that time it has become a useful tool in the clinician’s armamentarium. Extended TIPS with or without thrombolysis can be used in cases of BCS coupled with portal vein thrombosis (30). Two series with a total of 34 patients showed improved patency and less dysfunction with polytetrafluoroethylene (PTFE)-covered stents compared with bare stents (35, 36). Other series using TIPS in BCS report eventual transplant in 10–40% of patients (16). Misplacement or migration of TIPS into the portal vein is not uncommon (Fig. 5) (9, 37). This particular complication can have adverse consequences if the patient goes on for transplantation.

A case series of 61 patients who underwent TIPS and/or hepatic vein recanalization with angioplasty and stenting showed excellent survival. However, 40

Fig. 4. Stent placement (arrow) across an inferior vena cava (IVC) stenosis in a patient with Budd–Chiari syndrome (BCS).

Fig. 5. Coronal magnetic resonance imaging (MRI) showing (a) cranial migration of transjugular intrahepatic portosystemic shunt (TIPS) (arrow) and resulting impingement on the inferior vena cava (IVC). (b) The impingement of the IVC was relieved by placing a stent (white arrow) in the IVC adjacent to the TIPS (black arrow).
patients required repeated radiological interventions. An important conclusion from this study was that short-length stenosis of hepatic veins is under-recognized and percutaneous recanalization is under-utilized in the management of patients with BCS. This study suggested that when stratified by disease severity, interventional radiology techniques may have the most impact on the sickest patients (compared with survival in patients treated with surgical shunts). Forty-four patients treated initially with surgical shunts, transplantation or medical therapy alone were not included for analysis in this study (26).

Interpretation of the literature in this area can be difficult. Results from a case series in Asia may not apply to the patient population in Western countries because the incidence of membranous obstruction is much higher in the Asian series. Also, technology evolves quickly and today’s stents used for TIPS and other percutaneous interventions may have very different properties than those from only a few years ago. As stated before, no prospective trials in patients with BCS exist and conclusions from any published report should be viewed with caution.

**Surgical shunts**

The role of surgical shunts in the treatment of BCS is a controversial topic. A wide range of perioperative mortality has been reported (0–50%) (3). However, long-term survival rates (5–14 years) after various shunting procedures have been reported to be as high as 90% (16). There are multiple factors that make it difficult to interpret the literature on this subject. Studies that compare surgical shunts to medical or minimally invasive techniques are often difficult to interpret because they are usually retrospective, may have significant time bias and, maybe most importantly, the sickest patients are usually managed non-operatively (38). Also, in recent years, experience with these procedures has begun to dwindle. For example, chief residents at a large academic centre such as Johns Hopkins only performed four portosystemic decompression surgeries between 1995 and 1999 (9).

Shunting procedures are indicated in patients with reversible liver injury. However, identifying this population of patients is not straightforward (39). Anatomic considerations such as extensive IVC thrombus may make TIPS difficult and favour placement of a surgical shunt. While several cases of BCS with portal vein and/or mesenteric vein thrombosis have been successfully managed with TIPS and thrombolysis (30), open surgical intervention may be required.

Selection of which surgical shunt to perform requires consideration of several factors. For patients without IVC obstruction, the portocaval or mesocaval shunts are reasonable options. Previous portocaval shunts can make the hilar dissection during transplantation particularly challenging. In patients with caudate lobe hypertrophy (Fig. 1), portocaval shunts may be difficult and even require caudate lobe resection before construction (9). Orloff et al. (40) reported great success with portocaval shunts with 31 of 32 patients alive at 3.5–27 years after surgery. Unfortunately, their success has not been replicated by others (9).

For patients with IVC obstruction and/or an infrahepatic to right atrium pressure gradient of > 20 mm Hg, mesocaval and portocaval shunts may not effectively decompress the liver. Some authors report improved long-term outcomes with a pressure gradient of greater than 10 mm Hg between the portal vein and IVC (40). The most important pressure gradient is not between the infra and suprahepatic vena cava, but between the two vascular structures that are to be connected (3). Consequently, many authors consider mesoatrial or cavoatrial shunts in these cases (Fig. 6) (9). While the primary patency rate of mesoatrial shunts is relatively low, 5-year survival has been acceptable (9, 41).

Wang (2) reported good 1-, 3- and 5-year patency rates for mesoatrial (90.7%, 77.1%, 61.1%) and cavoatrial (97.2%, 86.0%, 79.8%) shunts. Orloff reported 18 cases of BCS with IVC and hepatic vein occlusion. In their early experience, eight of these patients were treated with mesoatrial shunts. Five of these patients died after the graft thrombosed. The
The first successful liver transplant for a patient with BCS was performed in 1974 (46). Since that time, several series and registry analyses of liver transplantation for BCS have been reported (Table 1). Recently, two large series have been published documenting the European and U.S. experience (47, 48). For patients with BCS and acute fulminant liver failure or cirrhosis and decompensated disease with poor synthetic function, liver transplantation is the treatment of choice (9). Evidence of poor hepatocyte synthetic function includes albumin <3 g/dl, prothrombin time 3 s greater than control, and conjugated bilirubin >3 mg/dl (23). When compared with patients who receive shunting procedures, transplanted patients maintain higher serum albumin levels and improved synthetic function (25). In patients with certain synthetic deficiencies such as antithrombin III or protein C, liver transplantation offers the additional advantage of curing these genetic defects (9, 49, 50).

While liver transplantation has emerged as a safe therapeutic option for patients with BCS, its application should not be used liberally. Patients with BCS have multiple therapeutic options that may not be available to many patients on the transplant waiting list.

Calculating model for end-stage liver disease (MELD) scores in patients with chronic BCS may be problematic because many of these patients are on warfarin or some other form of anticoagulation. In this patient population, international normalized ratio (INR) is not an accurate indicator of hepatic synthetic function. It has recently been suggested that the INR be capped at 2.5 for patients on warfarin. Additionally, it has been proposed that patients with fulminant hepatic failure resulting from BCS should be listed as United Network for Organ Sharing (UNOS) status 1A (51).

Several technical considerations make the transplantation procedure itself especially challenging in patients with BCS. Haemostasis can be particularly challenging, and above average blood loss is often reported (39). Some centres employ venovenous bypass during all transplants in patients who have significant portal hypertension and prior shunt surgery. Caudate lobe enlargement (Fig. 1) may present problems during transplantation by making mobilization of the liver difficult and increasing the difficulty of the piggyback technique (9). BCS may cause a diffuse fibrotic reaction in the retroperitoneum, which may increase the complexity of steps requiring control of the IVC (9, 38). Additionally, dense adhesions between the liver and diaphragm surrounding the suprahepatic IVC have been described (38, 52). The portal vein may be thickened, narrowed and even thrombosed (39).
Segev published the largest series of transplantation for BCS (48). This study included 510 patients who were divided into three time periods: historical (1987–1997), pre-MELD (1998–2002) and MELD (2002–2006). The analysis found that the demographic profile of patients receiving transplant changed through time. When compared with previous eras, the MELD era recipients were less likely to be female, hospitalized, on life support or have prior transplants.

In the MELD era, a significant percentage of BCS patients (19%) received exception points, which caused an increase in the listing MELD compared with the calculated MELD (25.6 vs. 23.1 respectively). Graft and patient survival was significantly higher in the MELD era (Table 1). The study also evaluated the impact of preoperative TIPS on outcomes of liver transplantation. The study found that TIPS did not adversely affect graft loss or death over all time periods. The final analysis found that only life support, prior transplantation and prolonged cold ischaemia time (>12 h) were independent predictors of both graft loss and death. Taking into account the fact that MELD era patients were less likely to be on life support or have prior transplants, the improved patient survival over time may be a by-product of transplanting patients with less disease severity. The study did not calculate Child–Turcotte–Pugh (CTP) and MELD scores for the pre-MELD and historical groups, so direct comparison of disease severity between the groups is problematic. As opposed to the European experience, patients in the MELD era did not suffer inordinate early deaths or graft loss. Owing to the limitations of the UNOS database, the underlying aetiology of BCS in this cohort could not be captured.

From 1968 to 2001, the European Liver Transplant Registry (ELTR) recorded 391 patients who received transplantation for BCS with a 1-, 5- and 10-year patient survival of 73%, 68% and 63% respectively. Overall 1-, 5- and 10-year patient survival for all indications during this time period was 83%, 71% and 63% respectively (53). The cohort of patients with BCS represented 1% or all transplants during that time period. The important observation here is that short-term survival in patients with BCS is lower while long-term survival is equivalent.

Mentha presented a cohort of patients, 256, undergoing transplantation for BCS in Europe. Multivariate analysis found the only pretransplant predictors of mortality to be impaired renal function (RR = 3) and a history of a surgical shunt/TIPS (RR = 2.2). However, the authors warned against interpreting this result to mean that surgical shunts or TIPS should be abandoned because, by definition, this cohort only included patients whose shunts had failed. The need

### Table 1. Liver transplantation for Budd–Chiari syndrome

<table>
<thead>
<tr>
<th>Author</th>
<th>Year</th>
<th>Country</th>
<th>Years included</th>
<th># patients</th>
<th>PreTx shunt</th>
<th>PreTx TIPS</th>
<th>1-year patient survival (%)</th>
<th>3-year patient survival (%)</th>
<th>5-year patient survival (%)</th>
<th>Recurrence</th>
<th>% retransplant</th>
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<td>1976–1986</td>
<td>17</td>
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<td>88</td>
<td>88</td>
<td>NS</td>
<td>1</td>
<td>12</td>
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<tr>
<td>Jamieson</td>
<td>1991</td>
<td>UK</td>
<td>1986–1990</td>
<td>26</td>
<td>3</td>
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<tr>
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<td>1984–1990</td>
<td>14</td>
<td>2</td>
<td>NS</td>
<td>86</td>
<td>76</td>
<td>76</td>
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<td>1988–1994</td>
<td>8</td>
<td>NS</td>
<td>NS</td>
<td>88</td>
<td>NS</td>
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<td>88</td>
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<td>1997–2004</td>
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<td>9</td>
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<td>Segev</td>
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<td>23</td>
<td>88</td>
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\(^1\)All procedures were live donor liver transplants.

\(^*\)Time period only includes patients from the MELD era.

NS, not stated; MELDS, model for end-stage liver disease; TIPS, transjugular intrahepatic portosystemic shunt.
for emergency transplant was not associated with increased mortality. Eighty-seven per cent of deaths occurred during the first year and the authors concluded that the slow course of myeloproliferative disorders was not significantly affected by transplantation (47). However, two other series report cases of polycythaemia vera transforming into acute leukaemia several years after transplant (49, 54). The life expectancy for patients with polycythaemia who have developed myelofibrosis is approximately 3 years. Thus, transplantation does not offer a survival advantage in patients with myelofibrosis (54). Based on Murad’s classification system of patients with BCS, patients in class III from Mentha’s cohort had improved survival with transplantation compared with Murad’s cohort treated without transplant (5-year survival 71% vs. 42%) (44, 47).

Yamada presented nine patients with BCS treated with living donor liver transplantation. This series reported an 88% 1-year and 71% 3-year survival, with two patients suffering recurrence of BCS (55).

Liver transplants complicated by postoperative BCS may not always be amenable to minimal invasive interventions (34). In a review of 1112 patients who underwent liver transplantation with the piggyback technique, nine patients suffered from early postoperative BCS. Retransplantation was performed in seven of these patients, and four of the nine died within 2 months of their operation (7). Other large series have reported a 24% mortality rate for this complication (6). Other than retransplantation and percutaneous interventions, alternative management strategies include surgical shunts (56), simple rotation of the graft, caval thrombectomy, anastomosis reconstruction or diaphragmatic placement of the graft (6).

Budd–Chiari syndrome following liver transplantation with the piggyback technique is a well-described complication with a reported incidence of 1–7% (8). Venous outflow obstruction is often considered more common after the piggyback technique (33). However, two recent retrospective studies were unable to detect a difference in the rate of BCS between the piggyback technique and the standard technique (57, 58). Furthermore, there is little difference in postoperative BCS when comparing large case series utilizing either the standard or piggyback technique, 1.7% vs. 1.1–1.5% respectively (6, 7, 59). This complication usually presents in the early postoperative period. However, approximately 25% of cases may present after the first postoperative week (7). Technical factors that have been associated with this complication are inadequate graft size and use of two hepatic veins for the venous anastomosis (6, 7). The rate of BCS after construction of the venous anastomosis with two veins is 2.3% while the use of three veins decreases the rate to 0.7% (7). Compared with the conventional piggyback technique, the side-to-side variation of the piggyback technique has been associated with a decreased incidence of BCS, 2.4% vs. 0.7% respectively ($P = 0.014$) (6).

As recurrent BCS after transplantation has been reported, lifelong anticoagulation is recommended (4). Recurrence of BCS after transplantation has been reported to be as high as 27% and may require retransplantation (49). Patients are often started on heparin postoperatively (total daily dose 7500–30 000 U intravenously) and converted to coumadin a week later (23, 54, 60). In patients with myeloproliferative disorders, coumadin does not address their underlying pathophysiology. In these patients, administration of hydroxyurea and aspirin can be given instead of traditional anticoagulation to treat the aetiology of BCS. The newer drug, anagrelide, has replaced hydroxyurea in some centres (23). Posttransplant anticoagulation may not be necessary in patients who possessed synthetic defects such as protein C deficiency corrected by transplantation (23, 50). While life-long anticoagulation after transplantation is routine for most patients, it does come with a price. Campbell (47) and Mentha (52) reported clinically significant haemorrhage in many patients, 44% and 11% respectively.

**Prognosis**

Outcomes in patients with BCS have improved since 1985 (19). Approximately 25% of patients remain asymptomatic after treatment, and spontaneous resolution has been reported (16). Several authors have investigated prognostic indicators in patients with BCS.

Khuroo (29) analysed 47 consecutive patients with BCS and found the following factors to adversely affect survival: florid clinical presentation, male sex, no TIPS performed and CTP score.

Langlet updated previous work on their prognostic index (PI) and found that ascites score, Pugh score, age, creatinine and type III presentation (acute injury superimposed on chronic lesions) were all independent predictors of survival. After including 123 patients who underwent surgical shunts, this analysis was unable to show a survival benefit to surgical shunts whether performed early (within 2 months after diagnosis) or not. Short-term and long-term survival was good regardless of treatment modality in patients with PI < 5.1. However, the authors caution
Table 2. Murad's Budd–Chiari syndrome prognostic classification

<table>
<thead>
<tr>
<th>Class</th>
<th>Score</th>
<th>% of cohort (%)</th>
<th>5-year survival (%)</th>
<th>% with surgical shunt</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>0–1.1</td>
<td>27</td>
<td>89</td>
<td>29</td>
</tr>
<tr>
<td>II</td>
<td>1.1–1.5</td>
<td>46</td>
<td>74</td>
<td>55</td>
</tr>
<tr>
<td>III</td>
<td>&gt; 1.5</td>
<td>27</td>
<td>42</td>
<td>60</td>
</tr>
</tbody>
</table>

*Equation: 1.27 × encephalopathy + 1.04 × ascites + 0.72 × prothrombin time + 0.004 × bilirubin.

Ascites and hepatic encephalopathy were scored as present (1) or absent (0) and prothrombin time as higher (1) or lower (0) than 2.3 INR. Bilirubin was included as a continuous variable.

INR, international normalized ratio.

that this study was underpowered to detect a possible benefit to surgical shunts in patients who present with type I BCS (acute injury only, corresponding to the onset of hepatic outflow obstruction) (19). A weakness of the PI is that only two of its variables incorporate objective data.

Murad reported the results of an international multi-institutional study, which included 237 patients (205 included in multivariate analysis) treated with a variety of modalities. This cohort achieved transplant-free survival rates of 82%, 69% and 62% at 1, 5 and 10 years respectively. The following factors were found by multivariate analysis to be independent predictors of 5-year transplant-free survival: presence of ascites, presence of encephalopathy, INR and bilirubin. Based on these variables, the authors constructed an equation that assigned patients into one of three classes (Table 2). The 5-year transplant-free survival was 89%, 74% and 42% for class I, class II and class III respectively (44). The strengths of this analysis were that it included more patients than the Langlet study, and its variables were either objective or binary. The degree of ascites and encephalopathy were not variables. Only the presence or absence of ascites and encephalopathy was considered in the multivariate analysis. A possible weakness of this study is that it excluded patients who underwent transplant.

Conclusion

Budd–Chiari syndrome is a complex disease with a wide spectrum of aetiologies and presentations. A multidisciplinary approach to diagnosis and treatment is advantageous in patients with BCS. Medical therapy consists of treatment of underlying disease, anticoagulation and symptom control. Emerging technologies have offered new minimally invasive treatment modalities such as percutaneous catheter-directed thrombolysis, angioplasty, stenting and TIPS. While surgical shunting continues to play a role in the treatment of patients with BCS, its place in treatment strategies is unclear. Surgical shunting is best reserved to those institutions with significant operative experience, TIPS capability and a liver transplant programme. Liver transplantation is the treatment of choice for patients with fulminant hepatic failure or those with chronic cirrhosis and poor synthetic function. As technology continues to progress, new diagnostic and therapeutic options will likely enhance the clinician’s ability to care for patients with this challenging disease.

References


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