Review

DOI: 10.5582/irdr.2014.01031

Budd-Chiari syndrome and liver transplantation

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Summary **Budd-Chiari syndrome involves obstruction of hepatic venous outflow tracts at various levels from small hepatic veins to the inferior vena cava and is the result of thrombosis or its fibrous sequelae. There is a conspicuous difference in its etiology in the West and the East. Myeloproliferative disease predominates in the West and obstruction of the vena cava predominates in the East. The clinical presentation and clinical manifestations are so varied that it should be suspected in any patient with acute or chronic liver dysfunction. It should be treated with step-wise management. First-line therapy should be anticoagulation with medical treatment of the underlying illness, and interventional revascularization and TIPS are indicated in the event of a lack of response to medical therapy. Liver transplantation may be indicated as a rescue treatment or for fulminant cases with promising results. This step-by-step strategy has achieved a 5-year transplant-free survival rate of 70% and a 5-year overall survival rate of 90%. Living donor liver transplantation can also be used for patients with Budd-Chiari syndrome if deceased donor livers are scarce, but it requires a difficult procedure particularly with regard to venous outflow reconstruction.**

Keywords: Budd-Chiari syndrome, liver transplantation, deceased donor, living donor

1. Introduction

Budd-Chiari syndrome (BCS) is a rare disease with a multifactorial etiology and is characterized by obstruction of the hepatic venous outflow anywhere from the intrahepatic venules to the suprahepatic portion of the inferior vena cava (IVC). Regardless of its etiology, the subsequent increase in hepatic sinusoidal pressure will lead to portal hypertension and related clinical sequelae (*1*).

BCS varies greatly in terms of its etiology, clinical presentation, and management. The clinical presentation may be asymptomatic, chronic, or fulminant. The treatment strategy varies from medical anticoagulant and antithrombotic treatment to surgical therapy including liver transplantation (*2-4*).

Released online in J-STAGE as advance publication January 5, 2015.

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The aim of this review was to encompass the updated practical management of this disease entity with special reference to liver transplantation.

2. Etiology and epidemiology

BCS is not a primary disease of the liver parenchyma but subsequent liver dysfunction following obstruction of hepatic veins or the suprahepatic IVC. Hepatic venous outflow obstruction results in an elevated sinusoidal pressure and leads to hepatic congestion. Usually, congestion is followed by subsequent centrilobular fibrosis and nodular regenerative hyperplasia that lead to chronic liver dysfunction and cirrhosis; in some instances, however, it results in fulminant hepatic failure requiring emergency liver transplantation (*5*).

There is an interesting but not as yet understood difference in the etiology and epidemiology of this condition in the West and East (*6*). Recent studies from Western countries have revealed that primary BCS can be regarded as a multifactorial disease in which several prothrombotic conditions additively predispose patients to develop thrombosis in hepatic veins (*3,7,8*). Common prothrombotic conditions associated with BCS include inherited and acquired hypercoagulable states.

Inherited conditions such as factor V Linden mutation (*9,10*), protein C/S deficiency (*3,11,12*), antithrombin III deficiency (*12*), and the prothrombin G20210A mutation (*13*) are common causes of hepatic venous thrombosis resulting in BCS. Acquired hypercoagulable condition such as myeloproliferative disorders (MPD) including polycythemia vera, paroxysmal nocturnal hemoglobinuria, essential thrombocytosis, agnogenic myeloid metaplasia, and myelofibrosis account for 30% to 50% of BCS cases in Western countries (*8*). Recently, a particular somatic mutation (V617F) in the Janus tyrosine kinase-2 (JAK2) gene in myeloid cells was identified in a study of chronic MPD (*14*). This mutation has been detected in around 40% of patients with primary BCS (*15-17*). Other causes of BCS such as Behcet's disease (*18,19*), antiphospholipid syndrome (*20*), and oral contraceptives (*21*) have been reported.

In Western countries, a prothrombotic condition leading to hepatic venous thrombosis is most often the cause of BCS, while in Eastern countries BCS is most often caused by the membranous obstruction of the vena cava (MOVC) or primary IVC thrombosis (*6,22*). Reports from China (*23*), Nepal (*24*), India (*25*), and South Africa (*26*) have revealed that almost all cases of BCS were caused by MOVC. This has also been the case in Japan, although hypercoagulable patients with hepatic vein thrombosis appear to be increasing (*27*). The reason for these epidemiological discrepancies has yet to be fully elucidated, but a possible explanation may be that thrombophilic genetic changes are more common among Caucasians and thus result mostly in hepatic venous thrombosis, while some infections are more common in Eastern countries that predispose Asians to IVC thrombosis (*6,28,29*).

3. Clinical presentation, diagnosis, and prognosis

The clinical presentation of BCS may be fulminant (5%), acute (20%), or subacute/chronic (60%) (*5*), though 15-20% of patients with BCS may be asymptomatic (*30*). Although nonspecific, the triad of hepatomegaly, abdominal pain, and ascites is typically present in patients with BCS. Lower extremity edema is also frequently encountered. Nausea, vomiting, and jaundice can develop more often in patients with fulminant or acute forms of BCS. In contrast, findings associated with portal hypertension such as splenomegaly, esophageal varices, encephalopathy, and gastrointestinal bleeding are more commonly seen in the chronic form. In some patients, the hepatic sinusoids may be completely decompressed *via* large portosystemic and intrahepatic collaterals so that they are completely asymptomatic (*31*).

Fulminant BCS: Fulminant BCS develops within a few days, presenting as acute liver failure with elevated liver enzymes, hyperbilirubinemia, coagulopathy, and encephalopathy (*32*). The liver is severely enlarged, with massive ascites and acute renal failure. This is an absolute indication for liver transplantation.

Acute BCS: Acute BCS develops usually within 1 month and is characterized by intractable ascites, abdominal pain, liver enlargement, renal failure, elevated liver function test results, and coagulopathy (*33*).

Subacute BCS: Subacute BCS is the most common clinical form of BCS in prothrombotic patients. This form of BCS has an insidious onset and may take as long as 3 months to become asymptomatic with the development of decompressive collaterals. Treatment should be started during the subacute phase before BCS becomes chronic (*33*).

Chronic BCS: This form is characterized by the development of portal hypertension. There is marked abdominal distension due to massive ascites, while liver function test results are minimally affected or normal. Renal failure is seen in 50% of patients and esophageal variceal bleeding is encountered in 15-20% of these patients (*33*).

3.1. *Diagnosis*

Physicians should always keep BCS in mind when seeing patients with acute or chronic liver disease. BCS is more likely when there is no other, more common, cause of liver disease or when there is a known underlying prothrombotic condition. The essential point is to consider BCS in patients with known prothrombotic states who present with abnormal liver function test results, upper abdominal pain, and ascites.

Definitive diagnosis is based on evidence of an obstructed hepatic outflow including that in the hepatic veins or suprahepatic IVC, and in principle, is reached based on findings of dilation of the hepatic veins upstream of an obstacle, the presence of a thrombus in the hepatic veins or IVC, the transformation of the veins into a cord devoid of flow signal, and venous collaterals depicted as an abnormally enhanced vessels branching to or from the hepatic veins or IVC. Doppler ultrasonography, contrast-enhanced triphasic computed tomography, and magnetic resonance imaging are sufficient to reveal these diagnostic features of BCS (*31,34*). Nowadays, there is no call for direct or retrograde venography solely to diagnose BCS, but patients with BCS are usually referred to an interventional radiologist to confirm the diagnosis and for therapeutic interventions (*35*). Catheter venography is considered the reference standard for the diagnosis of BCS. It provides anatomical information by directly depicting venous problems, hemodynamic and venous pressure measurements, and histologic information by facilitating a transjugular liver biopsy and it allows the possibility of endovascular management of the outflow obstruction.

3.2. *Prognosis*

Incidentally found, asymptomatic forms of BCS have

a good prognosis (*30*), but symptomatic forms have a poor spontaneous course; an estimated 90% of untreated patients die within three years (*36*). Circumstances have changed with advances in the management of BCS, allowing a 5-year survival rate on the order of 90% (*4*). A step-wise management strategy aimed at minimal invasiveness is recommended by expert panels is based on the response to previous therapy rather than on the actual severity of the condition, and this strategy has considerably improved the survival expectancy and the quality of life for patients with BCS (*2,37,38*). Several prognostic indices (PI) have been evaluated to predict outcomes for patients with BCS such as the Child-Pugh score (*39*), model for end-stage liver disease (MELD) score (*40*), Clichy PI (*41,42*), Rotterdam BCS PI (*43*), and BCS-TIPS PI (*44*). The BCS-specific PI, Rotterdam PI, and BCS-TIPS PI appear to be preferable for clinical studies but insufficient for individual management (*37,45*).

4. Treatment

Therapeutic approaches to treat BCS are diverse and should be adapted depending on disease severity. Recently, an expert panel advocated a step-wise approach to treating BCS; 1. Anticoagulation, 2. Angioplasty and stenting, 3. TIPS or surgical shunt, and 4. Liver transplantation (*2*). A recent report from Europe (*37*) revealed that this step-wise management has improved the 5-year patient survival to 72% and transplant-free survival to 72%.

4.1. *Anticoagulation therapy*

Logically, prompt recognition and initiation of treatment of the underlying disease is recommended when BCS is diagnosed, and anticoagulation therapy should immediately be started even for asymptomatic patients (*46*). Although specific therapy for underlying prothrombotic disease is crucial, this aspect is beyond the scope of this article. Low molecular weight heparin for the initiation of anticoagulation therapy and subsequent long-term anticoagulation with warfarin to achieve an international normalized ratio for prothrombin time of 2.0 to 2.5 are recommended (*2*).

4.2. *Angioplasty and stenting*

Catheter-directed thrombolytic therapy, angioplasty, and stent placement may be effective in treating acute BCS. Thrombolytic therapy is considered for patients with the acute form of BCS and especially in rare situations where angiography reveals a fresh thrombus. Urokinase (240,000U per hour for 2 hours, followed by 60,000U per hour) or tissue plasminogen activator (0.5 to 1mg per hour) is infused directly into the thrombosed hepatic vein for about 24 hours *via* an inserted catheter (*1,47*). Percutaneous or transhepatic angioplasty of localized segments of the narrowed hepatic vein or IVC membranous obstruction may relieve symptoms in more than 70 percent of patients (*48,49*). Short stenosis either of the hepatic veins or of the IVC is found in about a third of patients, and the restoration of outflow through just one of the three main hepatic veins is usually sufficient to relieve symptoms (*50*) . Stent insertion may be considered if there is an inadequate response to balloon angioplasty or it may be reserved for cases of recurrent stenosis or occlusion. Unfortunately, however, angioplasty in combination with anticoagulation therapy has been reported to successfully control BCS in only 20-30% of patients, at least in reports from Western countries where hepatic vein thrombosis predominates (*38*). In contrast, a Chinese report of 115 patients noted success rates of 94% and 87% for stents placed in the IVC and hepatic veins, respectively (*51*).

4.3. *Transjugular intrahepatic portosystemic shunt (TIPS) and surgical shunts*

In patients with BCS that is not fully controlled by the aforementioned treatments, the next step is either TIPS or a surgical shunt.

For patients presenting weeks to months after hepatic vein thrombosis, the obstruction is generally no longer amenable to thrombolysis or angioplasty. TIPS is recommended as the next step in management. TIPS is useful in patients with an occluded IVC, those in whom the portal vein-IVC pressure gradient is less than 10 mmHg, and those with poor liver function reserve. TIPS is also recommended for those with the acute form of BCS who failed to respond to thrombolytic therapy (*2,37*). TIPS is the most common intervention for BCS in Europe, and many studies have reported its high success rate and relatively low rate of complications (*52-55*). Compared to an open surgical shunt, TIPS is associated with lower morbidity and mortality (*56,57*), but its drawback is frequent shunt occlusions requiring repeated interventions. The development of covered stents, however, has significantly improved the patency of TIPS in BCS (*53,54*). Rossle *et al*. (*58*) achieved initial success in 33 of 35 patients with 1- and 5-year transplant-free survival rates of 93% and 74%, respectively. In another study, TIPS was successfully performed in 124 of 133 patients with a clinical success rate of 84% (*44*). A recent European multicenter study reported a 5-year transplant-free survival rate of 72% in 157 patients with BCS who were treated with TIPS (*37*).

A surgical portosystemic shunt is recommended for patients with the subacute form of BCS when the underlying disease is associated with a favorable long-term outcome, patients have preserved liver function (Child-Pugh class A), and a liver biopsy reveals ongoing hepatic necrosis (*59*). A pressure gradient between the portal vein and IVC of more than 10 mmHg is

associated with a successful long-term outcome. Surgical shunts include a side-to-side portocaval shunt, a central splenorenal shunt, and a mesocaval shunt. The 5-year survival rate after surgical shunting ranges from 75% to 94%, with the higher end of the range being achieved when the IVC is not occluded (*60,61*). The essential aspect is to use a side-to-side portocaval shunt in the early stage of BCS to achieve an excellent outcome for patients with BCS (*59,62*). The rationale for surgical portosystemic shunting is to convert the portal flow into an outflow tract of the liver, and some patients with the severe form of BCS may potentially benefit from this procedure. However, no studies have described the survival benefit of surgical shunts (*42*). In light of advances in the TIPS procedure and accumulated evidence showing the impact of TIPS on patient survival, TIPS is preferred as the first choice for safe and optimal decompression.

5. Liver transplantation for BCS

In the remaining 10% to 20% of patients with BCS treated with a step-wise management strategy, anticoagulation, angioplasty, and TIPS fail either due to technical failure or to poor clinical results of a technically successful procedure resulting in the need for rescue transplantaion. Liver transplantation may also be the treatment of choice in patients with fulminant liver failure and those with highly advanced liver cirrhosis (*3*).

5.1. *Deceased donor liver transplantation for BCS in Western countries*

A search of the literature indicated that more than 1000 patients with BCS have undergone liver transplantation. Table 1 summarizes recent reports of liver transplant for a considerable number (> 10) of patients with BCS

Table 1. Deceased donor liver transplantation for BCS in Western countries

Author	Year	Country	Period studied	*n*	Etiology (*n*)	Indication (*n*)	Shunt *n* (%)	Follow-up	90-day mortality	Survival
Srinvasan *et al*. (*63*)	2002	UK	1988-1999	19	MPD (11) PD (2) Others (6)	Advanced cirrhosis (13) Acute liver failure (6)	5 (26%)	1-119 months (median 89)	5%	95% (1 year) 95% (3 years) 95% (5 years)
Cruz *et al*. (*64*)	2005	US	1988-2002	11	MPD (8) PD (1) Others (2)	Advanced cirrhosis (8) Acute liver failure (3)	5 (45%)	1-132 months (median 56)	11%	81% (1 year) 65% (5 years) 65% (10 years)
Ulrich *et al*. (*65*)	2008	Germany	1988-2006	42	MPD (13) PD (11) Others (18)	Advanced cirrhosis (22) Acute liver failure (20)	10 (24%)	1-203 months (median 96)	7%	92% (1 year) 89% (5 years) 84% (10 years)
Chinnakotla *et al*. (*66*)	2011	US	1987-2007	25	MPD (15) PD (6) Others (4)	Advanced cirrhosis (23) Acute liver failure (2)	3 (12%)	7-264 months (median 96)	4%	92% (1 year) 88% (5 years) 72% (10 years)
Mackiewicz *et al*. (*67*)	2012	Poland	2000-2009	24	MPD (3) PD (7) Others (14)	The Advanced cirrhosis (18) Acute liver failure (6)	6 (25%)	NA	13%	80% (1 year) 80% (3 years) 80% (5 years)
Seijo *et al*. (*37*)	2013	Europe Multicenter	2003-2009	20	NA	Advanced cirrhosis (6) Acute liver failure (14)	6 (30%)	0.1-74 months (median 50)	NA	95% (1 year) 89% (3 years) 78% (5 years)
Registry study										
Mentha *et al*. (*68*)	2006	Europe Registry	1988-1999	248	MPD (116) PD (71) Others (61)	Advanced cirrhosis (136) Acute liver failure (50)	57 (23%)	Median 48 months	21%	76% (1 year) 72% (5 years) 68% (10 years)
Segev *et al*. (*69*)	2007	US Registry	1987-2006	510	NA	Unknown (62) NA	NA	NA	15%	82% (1 year) 76% (3 years)

MPD, myeloproliferative disorder; PD, Prothrombotic disease; NA, not available

Table 2. Living donor liver transplantation for BCS in Asian countries

Author	Year	Country	Type of report	Age/ Gender	Etiology	Indication	Shunt	Graft type	Outflow reconstruction	Outcome
Haberal *et al.* (*74*)	1999	Turkey	Case report	17/F	Unknown	AC	No	Left lobe	Auxiliary heterotopic partial liver transplantation	Alive, 4 months
Nezakatgoo *et al.* (*75*)	1999	Iran	Case report	14/M	Unknown	AC	No	Left lobe	NA	Alive, 2 months
Yamada *et al.* (*71*)	2006	Japan	Case series	3/M	MOVC	AC	No	Left lateral sector	IVC-Left hepatic vein, piggy-back	Alive, 15 years
				10/M	MOVC	AC	No	Left lobe	IVC patch plasty, piggy back	Alive, 7 years
				11/M	MOVC	AC	No	Left lobe	IVC-Left hepatic vein, piggy-back	Dead, 1 month
				11/M	PD	AC	No	Left lobe	IVC patch plasty, piggy back	Alive, 7 years
				26/M	MOVC	AC	Yes	Right lobe	IVC patch plasty, piggy back	Dead, 17 months
				27/M	MOVC	AC	No	Right lobe	IVC patch plasty, piggy back	Alive, 16 months
				39/M	MOVC	ALF	No	Right lobe	IVC-Right hepatic vein, piggy-back	Alive, 6 years
				32/F	MPD	AC	No	Right lobe	IVC patch plasty, piggy back	Alive, 4 years
				17/M	PD	AC	No	Left lobe	IVC-Left hepatic vein, piggy-back	Alive, 10 months
Yan *et al.* (*76*)	2006	China	Case report	35/M	MOVC	AC	Yes	Right lobe	IVC interposition with cryopreserved homologous IVC	Alive, 3 months
Shimoda *et al.* (*77*)	2007	Japan	Case report	40/F	MOVC	AC	Yes	Right lobe	IVC interposition with autologous veins	Alive,17 months
Liu *et al.* (*78*)	2008	Taiwan	Case report	11/M	MOVC	AC	No	Left lobe	IVC patch plasty, piggy back	Alive, 2 years
Sasaki *et al.* (*79*)	2009	Japan	Case report	10/M	MOVC	AC	Yes	Left lateral sector	IVC interposition with cryopreserved homologous IVC	Alive, 2 months
Kawaguchi *et al.* (*80*)	2009	Japan	Case report	20/F	MPD	AC	No	Right lobe	IVC-Right hepatic vein, piggy-back	Alive, 1 month
Kazimi *et al.* (*81*)	2009	Turkey	Case report	29/M	MOVC	AC	No	Right lobe	Direct anastomosis right atrium and right hepatic vein, end-to-end	Alive, 3 months
Choi *et al.* (*72*)	2010	Korea	Case series	44/F	MOVC	AC	No	Right lobe	IVC-Right hepatic vein, piggy-back	Alive, 2 years
				45/M	MOVC	AC	No	Right lobe	IVC-Right hepatic vein, piggy-back	Alive, 18 months
				50/F	MOVC	AC	No	Right lobe	IVC interposition with cryopreserved homologous IVC	Alive, 13 months
				41/F	MOVC	AC	Yes	Right lobe	PTFE graft interposition between right atrium and right hepatic vein	Alive, 2 years
Shirai *et al.* (*82*)	2011	Japan	Case report	26/M	MOVC	ALF	No	Left lobe	IVC-Left hepatic vein, piggy-back	Alive, 1 year
Ogura *et al.* (*83*)	2011	Japan	Case report	36/M	MOVC	AC	Yes	Right lobe	IVC interposition with PTFE graft	Alive, 2 years
Soyama *et al.* (*84*)	2011	Japan	Case report	63/M	Unknown	ALF	No	Right lobe	Thrombectomy, IVC-Right hepatic vein, piggy-back	Alive, 1 month
Iwasaki *et al.* (*85*)	2012	Japan	Case report	22/F	MPD suspected	AC	No	NA	NA	Alive, 4 years
Sakcak *et al.* (*86*)	2012	Turkey	Case report	12/F	Iatrogenic	AC	Yes	Left lateral sector	IVC interposition with cryopreserved homologous aorta	Alive, 4 months
Bas *et al.* (*73*)	2012	Turkey	Case series	34/M	MPD	AC	No	Right lobe	IVC-Right hepatic vein, piggy-back	Alive, 30 months
				27/F	MPD	AC	No	Right lobe	IVC-Right hepatic vein, piggy-back	Alive, 18 months
				25/F	MPD	AC	No	Right lobe	IVC-Right hepatic vein, piggy-back	Alive, 6 months
Fukuda *et al.* (*87*)	2013	Japan	Case report	34/F	MOVC	AC	Yes	Left lobe	Supraphrenic vena cava-Left hepatic vein, end-to-end	Alive, 5 years
The present report		Japan	Personal experience	43/M	Unknown	ALF	No	Right lobe	IVC patch plasty, piggy back	Alive, 10 years
				52/M	MOVC	AC	No	Right lobe	Interposition with cryopreserved homologous IVC between right atrium and right hepatic vein	Alive, 14 months

IVC, inferior vena cava; PTFE, polytetrafluoroethylene; MOVC, membranous obstruction of the vena cava; MPD, myeloproliferative disorder; PD, Prothrombotic disease; AC, Advanced cirrhosis; ALF, Acute liver failure; NA, not available.

(*37,63-69*). The early mortality rate in these reports ranged from 4% to 21%, and the 1- and 5-year survival rates ranged from 80% to 95% and from 65% to 95%, respectively. All of these outcomes seem acceptable in comparison to those for other diseases requiring liver transplantation. Recently, large retrospective database studies were reported in Europe (European Liver Transplantation Registry [ELTR]) and the United States (United Network for Organ Sharing [UNOS] registry). Segev *et al*. (*69*) examined the UNOS database of recipients who underwent liver transplantation for BCS (n = 510) from 1987 to 2006 and found that 1- and 3-year patient survival rates were 82% and 76%, respectively. When stratified based on use of MELD, patients treated during the days of MELD (n = 100, 3-year patient survival of 85%) had significantly better survival than those treated prior to MELD (n = 168, 3-year patient survival of 73%). A longer cold ischemic time (> 12 hr), preoperative life support, and retransplantation were found to be independent risk factors for poor patient survival, while preceding TIPS appeared to have no impact on patient prognosis. Representing the ELTR, Mentha *et al*. (*68*) reviewed 248 patients who underwent liver transplantation for BCS from 1988 to 1999. They reported overall 1-, 3-, and 5-year patient survival rates of 76%, 75%, and 72%, respectively. They found that renal failure and the presence of a shunt were independent predictors for patient survival.

5.2. *Living donor liver transplantation for BCS in Asian countries*

Due to the severe scarcity of deceased-donor liver grafts, living-donor liver transplantation (LDLT) has been the mainstay for patients with end-stage liver disease and acute liver failure, including BCS, in most Asian countries (*70*). A search of the English literature yielded 30 patients with BCS who underwent LDLT; all were from Asian countries. This literature search included 3 case series reports (*71-73*) and 14 case reports (*74-87*) (Table 2). Additionally, the current authors encountered two cases of BCS out of 535 cases of LDLT at the University of Tokyo Hospital, and these cases are also shown in Table 2. Most cases (19/32) were from Japan, six cases were from Turkey, four cases were from South Korea, one was from Iran, one was from China, and one was from Taiwan. The etiology of eighteen cases (60%) were MOVC, which was as expected. This indicates the common difference in epidemiology in the East and the West (*6*). A point worth mentioning is outflow reconstruction during LDLT for recipients with BCS; the deceased donor graft includes the hepatic IVC and hepatic veins, and the removal and replacement of the native hepatic IVC with a cavo-caval anastomosis between the recipient's native superior/inferior IVC and the donor IVC is easily accomplished. In contrast, the piggy back technique for LDLT involves the preservation of the recipient's IVC, and anastomosis between the recipient's IVC and graft hepatic vein is mandatory in the absence of the donor IVC. Thus, LDLT presents substantial challenges in terms of treating BCS. The key consideration for using LDLT to treat BCS is the management of a stenotic or occluded native IVC and the choice of techniques used to reconstruct the hepatic outflow. Many of the reports listed in Table 2 described venous reconstruction in various fashions, as briefly explained in the table. As in the literature, one of cases of MOVC that the current authors encountered required difficult outflow reconstruction. This was the interposition of the cryopreserved homologous IVC between the right atrium and the right hepatic vein. Replacement and interpositioning of the IVC with the vascular graft was done in 7 cases (22%), direct reconstruction of the outflow to the atrium (or supraphrenic IVC) was done in 4 (13%), and patch plasty of the IVC was required in 7 (22%).

Although the long-term outcomes of LDLT for patients with BCS could not be determined from the case report literatures, the Japanese Liver Transplant Society recently published a report on the nationwide LDLT registry in Japan. That report identified 41 cases of BCS. According to that report, the 1-, 3-, 5-, and 10-year cumulative patient survival rates after LDLT for BCS were 89%, 84%, 81%, and 81%, respectively (*88*).

6. Conclusion

BCS should be treated with a step by step treatment strategy. Physicians should be aware of the diverse etiology of BCS, and first-line therapy should be anticoagulation with medical treatment of the underlying illness (if indicated). Interventional revascularization and TIPS are indicated in the event of a lack of response to medical therapy. Liver transplantation may be indicated as a rescue treatment or for fulminant cases with promising results. LDLT can also be used for patients with BCS, but it involves a difficult procedure particularly with regard to venous outflow reconstruction.

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(Received December 10, 2014; Revised December 23, 2014; Accepted December 25, 2015)