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Hepatic Outflow Obstruction (Budd-Chiari Syndrome)

Experience with 177 Patients and a Review of the Literature

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Introduction

The Budd-Chiari syndrome (BCS) is characterized by structural and functional abnormalities of the liver caused by obstruction to the outflow of hepatic venous blood. Any part of the outflow tract, from the microscopic central veins to the right atrium of the heart, may be affected by a variety of pathogenic mechanisms leading to thrombosis. Traditionally, hepatic veno-occlusive disease (HVOD) (66) and cardiac conditions are included in the list of causes of BCS (55). However, their distinctly different etiologies and presentations justifies their consideration separately from classic BCS.

Initial studies of BCS described an acute, often dramatic disorder, characterized clinically by abdominal pain, ascites, hepatomegaly, and rapidly progressive hepatic failure. However, in the early years of this century, a more insidious presentation characterized by abdominal distension, portal hypertension, and tortuously engorged abdominal veins was also recognized (80). Patients with acute BCS dominated most reports from Western countries (14, 58, 70, 80), while those with chronic BCS constituted the majority reported from Japan (41, 48, 72, 75, 114), China (108, 109), South Africa (42, 91), and India (1, 25, 27, 45, 47, 104). The clinical course of patients varied from decades of relatively untroubled survival at one extreme end of the spectrum (38, 109), to death within a few days at the other (80). The etiology of BCS also differed between countries (58, 70, 80). In Western countries, primary myeloproliferative syndromes (58, 70, 80, 99), hypercoagulable states (70, 80, 100), steroidal contraceptives (58, 101), and tumors (70, 80) were responsible for most cases, while pregnancy (47), infections (1), and an inferior vena caval (IVC) web or membranous obstruction of the inferior vena cava (MOVC) (23, 25, 26, 41, 51, 72, 78, 84, 91, 109) were implicated in patients from Asian countries.

Initially, BCS was a pathologic curiosity. Most patients were either diagnosed preterminally or were studied in detail at autopsy (80). There was no effective treatment (80). However, the introduction of, first, invasive (14, 27, 36, 52, 70) and, later, noninvasive imaging techniques (8, 11, 12, 33, 34, 36, 43, 52, 59, 61, 67, 71, 74, 96) simplified the exact delineation of the vascular anatomy. Similarly, innovative surgical and interventional techniques have now provided effective long-term treatment (2, 39, 40, 50, 63, 65, 68, 90, 95, 107, 108, 113, 116, 117). There has thus been a justifiable recent resurgence of interest in this condition.

Budd-Chiari syndrome has, however, remained an uncommon disease, and most institutions have not had the opportunity to study a substantial number of patients (16, 40, 70). Two large reviews of the world literature, one spanning the years before 1959 (80) and the other covering the years between 1960 and 1980 (70), documented collected series of 164 and 243 patients, respectively. A recent study from China included 100 patients (109), as did another from South Africa (91).

This article summarizes our 25-year experience at this institute with 177 patients with radiologically confirmed BCS. This large cohort of patients allowed us to study the etiology, clinical manifestations, radiology, histology, and outcome of BCS resulting from a wide spectrum of diseases. The existing literature is sparse for pregnancy- and puerperium-related BCS, (47) as well as for childhood and adolescent BCS (35, 42, 98), but some patients with these disorders have been described in a series of articles published between 1970 and 1980 (25–27, 47) from our center.

Patients and Methods

All patients were studied in the Department of Hepatology, Postgraduate Institute of Medical Education and Research,

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Chandigarh, between 1967 and December 31, 1991. A tabulated record of the clinical presentation, physical findings, laboratory test results, radiologic findings, and outcome of treatment for each radiologically confirmed patient with BCS has been maintained in a special register. Care was taken to note the possible etiology, physical signs of portal hypertension, signs of IVC obstruction, and treatment. During the years of this study, over 10,000 patients with liver disease were examined in the liver unit. Of these, more than 2,000 had portal hypertension. Among the patients with portal hypertension at our center, 42% had cirrhosis, 36% had extrahepatic portal venous obstruction, 15% had non-cirrhotic portal fibrosis, and 7% had Budd-Chiari syndrome.

Every patient initially underwent either an IVC catheterization with contrast study (IVCC) or a percutaneous transhepatic hepatovenography (PTHV) as the primary invasive radiologic procedure. In the earlier part of the study, the complementary procedure was carried out only if indicated. Over the last decade, most patients have had both in one session. Standard techniques were used for both procedures (27). Manometric recording of IVC pressure was performed in several patients and a few had intrahepatic and portal system pressure recordings as well. Ultrasonography (US), computerized tomography (CT), and pulsed Doppler US became available only during the last few years. Currently, US and Doppler US are used as the initial screening test before invasive studies are performed.

Patients in whom radiologic proof of diagnosis was not available during life were excluded from this study (47). Excluded from this study were 10 patients with HVOD, 10 with neoplasia-related hepatic venous (HV) block demonstrated at autopsy, and 19 with amebic liver abscess and venous lesions at autopsy.

The data were analyzed with reference to the clinical presentation, etiology, natural course, vascular anatomy, and outcome. Patients with MOVC, pregnancy- or puerperium-related BCS, and onset during childhood (under 12 years) or adolescence (12–18 years) were also analyzed separately. Results were compared with those from patients from diverse geographic areas and with the largest published experience available.

Reports by Parker (80) and Mitchell et al (70) summarized the literature to 1980, and a MEDLARS search produced more recent information on etiology and treatment modalities. Abstracts from presentations at the Second International Symposium on BCS, Kyoto, Japan (October 1991) have also been used (33, 42, 44, 49, 75, 78, 106, 108, 112, 113, 116, 117).

Results

One hundred and seventy-seven patients, 96 males and 81 females (M:F 1.2:1), had radiologically confirmed BCS. Age at diagnosis ranged from 18 months to 70 years, while the duration of symptoms varied between 3 days and 22 years. Thirty-eight patients (21%) developed symptoms of BCS before the age of 18. Thirty-eight of the 81 females (47%) developed BCS in pregnancy or early puerperium.

One hundred and forty-four PTHV and 158 IVCC studies were performed on these patients. Fifteen patients underwent a definitive shunt procedure, 10 had transcardiac "finger fracture" of an IVC membrane, and 2 had a balloon membranotomy. Twenty-four patients died in hospital while a further 12 left in a moribund condition. As these patients were very unlikely to survive, they have been included in the mortality figures (Table 1).

Clinical presentation and etiology

Patients presented at this institution with symptoms of BCS of variable duration (Table 2). A distinct bimodal distribution was observed. Sixty-three patients (35.6%) came to hospital within 6 months of symptom onset, and two-thirds of these presented within the first 3 months. Almost two-thirds of all patients had been symptomatic for more than a year, and a significant number had been symptomatic for 5 and 10 years as well.

Twenty-nine patients were under 18 years of age at presentation (16%), while 13 (7%) were older than 50 years. The vast majority (77%) were between 19 and 49 years. There was no familial occurrence of BCS (32, 35).

In the entire group, complaints consisted of abdominal distension in 136 (77%), epigastric or right hypochondriac pain in 100 (57%), presence of distended abdominal veins in 72 (40%), upper gastrointestinal bleeding in 34 (19%), jaundice in 32 (18%), and hepatic encephalopathy in 20 (11%). The liver was enlarged in 159 patients (90%) and the spleen in 81 (46%). Free fluid was clinically detectable in 131 patients (74%), edema in 58

TABLE 1. Characteristics of 177 patients with Budd-Chiari syndrome

Number of patients	177
Male:Female	96:81 (1.2:1)
Mean age (range in years)	29.5 (1.5–70)
Mean duration of symptoms (range in months)	45.1 (0.1–264)
Total number of angiographic procedures	302
Surgical procedures performed	27 (15.2%)
Shunt operations	15
Hospital deaths	36 (20.4%)

TABLE 2. Duration of symptoms at presentation

Duration (mo.)	Parker (Ref. 80) N = 134 No. (%)	Present study N = 177 No. (%)	P
<6	91 (68.2)	63 (35.6)	<0.001
<1	26 (19.5)	17 (9.6)	
1–3	50 (37.6)	25 (14.1)	
4–6	15 (11.3)	21 (11.9)	
>6	43 (31.3)	114 (64.6)	<0.001
7–12	10 (7.5)	24 (13.6)	
7–9	—	8 (4.6)	
10–12	—	16 (9)	
>12	33 (24.8)	90 (51)	
13–59	—	46 (26)	
60–119	—	22 (12.4)	
>120	—	22 (12.4)	

(33%), and abdominal venous prominence in 128 (72%). Venous engorgement in the lumbar region, suggestive of IVC block, was noted in 95 patients (53%). The incidence of clinical manifestations in relation to the duration of symptoms has been summarized in Table 3.

The symptoms recorded in Table 3 appeared at varying times during the course of illness, which ranged from 3 days to 22 years. The presenting complaints, however, could be assigned to a few broad categories:

Acute fulminant BCS: With 7% of all patients, this group presented with severe pain, distension, jaundice, ascites, hepatomegaly, encephalopathy, and biochemical evidence of severe hepatocellular dysfunction. This form of BCS was most frequently encountered in the early puerperium and almost invariably ended fatally. The lag between onset and presentation was usually less than 4 weeks.

Acute BCS: With 28% of all patients, this group presented with pain, distension, and tender hepatomegaly with ascites. There was no encephalopathy, and many recovered. At presentation, these patients had been symptomatic for periods ranging from a few days to 6 months.

Chronic BCS: With 65% of all patients, this group presented with symptoms and signs of portal hypertension. They had large nodular livers, distended veins, and relatively preserved hepatocellular function despite the clinical diagnosis of cirrhosis. Variceal bleeding was common. About 20% of patients could recall an episode of acute BCS in the past. Some patients in this group presented with the solitary complaint of distended tortuous abdominal veins, without any clinical or biochemical features of hepatocellular dysfunction. The duration of symptoms ranged from 6 months to 22 years.

The transition from the acute to the chronic forms usually occurred between 7 and 12 months after onset of symptoms (Table 3).

We tried to establish a cause for BCS in all patients, but the study spanned a long period, and methods and their application were not uniform over time (82). Facilities for bone marrow culture and the study of hemopoietic cell lines were not available. Table 4 shows the relation between causes of BCS and duration of the disease. The group of pregnant or early puerperal patients was the easiest to diagnose, followed by the groups of those with tumors, hypercoagulable states, infections, and miscellaneous disorders. The latter included 2 patients each with abdominal trauma and postoperative dehydration, and 1 patient each with widespread neurofibromatosis, Behçet disease, sarcoidosis, and pulmonary hypertension. A large group of patients had a radiologically demonstrated IVC membrane or web, but there were also a substantial number in whom no cause could be found. While it was possible to accurately identify the cause in a majority of patients with short-duration BCS (46/63 patients; 73%), it was more difficult to establish a cause in patients with BCS of long duration (66/114 patients, 57%).

Our decision to use disease duration of less than

TABLE 3. Clinical features observed at presentation in 177 patients with Budd-Chiari syndrome in relation to duration of symptoms*

Duration (mo.)	<1	1–3	4–6	7–12	13–59	60–119	>120
No. pts.	17	25	21	24	46	22	22
Male:Female	1:3:2	2:3	2.5:1	2.5:1	1:1	2:1	1.5:1
Age at presentation (yrs.)							
<18	1 (6)	4 (16)	2 (10)	4 (17)	4† (10)	3 (14)	5 (23)
19–49	16 (94)	19 (76)	16 (76)	18 (75)	33 (82)	18 (82)	15 (68)
>50	—	2 (8)	3 (14)	2 (8)	3 (8)	1 (4)	2 (9)
Abdominal pain	14 (82)	21 (84)	15 (71)	6 (25)	24 (52)	11 (50)	9 (41)
Abdominal distension	16 (94)	22 (88)	20 (95)	16 (67)	31 (67)	17 (77)	14 (64)
Jaundice	6 (35)	10 (40)	7 (33)	1 (4)	4 (9)	1 (4)	3 (14)
Upper G.I. bleeding	1 (6)	2 (8)	1 (5)	2 (8)	13 (28)	7 (32)	8 (36)
Abdominal veins‡	1 (6)	—	—	10 (42)	28 (61)	16 (68)	17 (77)
Hepatic encephalopathy	6 (35)	7 (28)	—	—	5 (10)	1 (4)	1 (4)
Hepatomegaly	14 (82)	24 (96)	17 (81)	23 (96)	44 (96)	15 (68)	22 (100)
Splenomegaly	3 (18)	6 (24)	11 (52)	12 (50)	23 (50)	12 (55)	14 (64)
Ascites	17 (100)	23 (92)	19 (90)	15 (63)	29 (63)	15 (68)	13 (59)
Edema	5 (29)	11 (44)	5 (24)	6 (25)	12 (26)	11 (50)	8 (36)
Radiology (no. positive/total)							
Hepatic vein block (no./total)	15/15	22/23	17/17	17/23	30/40	11/15	7/11
Inferior vena cava block (no./total)	4/13	16/18	12/18	17/24	32/45	18/20	20/20

* Nos. in parentheses are percentages.

† No age available for 6 patients.

‡ These patients complained of abdominal venous distension as a symptom.

TABLE 4. Etiology of Budd-Chiari syndrome in relation to duration

Duration (mo.)	<1	1–3	4–6	7–12	13–59	60–119	>120	Total (%)
Cause								
Inferior vena cava membrane	0	6	4	9	13	9	13	54 (28.7)
Pregnancy	13	11	2	0	8	2	2	38 (20.2)
Tumors	3	1	0	2	0	1	4	11 (5.8)
Hypercoagulable states*	1	4	2	0	0	0	0	7 (3.7)
Infection	0	1	0	0	2	1	1	5 (2.6)
Others (see text)	0	0	1	0	4	3	0	8 (4.3)
Idiopathic	0	4	13	14	20	8	6	65 (34.6)

* Hypercoagulable states included 4 patients with polycythemia vera and 3 with paroxysmal nocturnal hemoglobinuria. Infections included 1 each amebic liver abscess and tuberculosis and 3 of hydatid disease. Six patients with inferior vena cava (IVC) membrane also had hepatocellular carcinoma and 5 patients with pregnancy-related Budd-Chiari syndrome had IVC membranes.

6 months to distinguish between acute and chronic BCS yielded a bimodal distribution. However, the relative proportion of those with acute and chronic BCS in our series was the reverse of that described by Parker (Table 2). A significant difference in clinical presentation emerged in the 2 forms (Table 5). Abdominal pain, distension, jaundice, hepatic encephalopathy, ascites, hyperbilirubinemia, pregnancy, and hypercoagulability were significantly more common in acute BCS, while upper gastrointestinal bleeding, splenomegaly, distended veins and MOVC were significantly more common in chronic BCS. Hepatomegaly was documented in most patients; however, some of the patients with acute BCS had ascites with tense abdominal distention, making measurement of the liver difficult. Hence, this group had a lower incidence of observed hepatomegaly.

TABLE 5. Acute and chronic Budd-Chiari syndrome: Clinical and etiologic differentiation*

		Acute (<6 mo.) N = 63	Chronic (>6 mo.) N = 114
Male:Female		1:12	1.5:1
Abdominal pain	($p < 0.001$)	50 (79)	50 (44)
Distension	($p < 0.001$)	58 (92)	78 (68)
Jaundice	($p < 0.001$)	23 (37)	9 (8)
Upper G.I. bleeding	($p < 0.005$)	4 (6)	30 (26)
Distended veins†	($p < 0.001$)	1 (2)	70 (61)
Hepatic encephalopathy	($p < 0.005$)	13 (20)	7 (6)
Hepatomegaly	(NS)	55 (87)	104 (91)
Splenomegaly	($p < 0.01$)	20 (31)	61 (54)
Ascites	($p < 0.001$)	59 (94)	72 (63)
Edema	(NS)	21 (33)	37 (32)
Serum bilirubin > 1 mg/dl	($p < 0.001$)	50 (77)	29 (25)
Serum albumin < 3 g/dl	(NS)	35 (55)	52 (47)
Hepatic vein block	(NS)	54/55	65/89
IVC block	(NS)	32/48	87/110
Pregnancy related	($p < 0.001$)	26 (41)	12 (11)
Hypercoagulability	($p < 0.001$)	7 (11)	0
IVC membrane	($p < 0.005$)	10 (16)	44 (39)
Tumors & others (see text)	(NS)	6 (9)	18 (16)
Idiopathic	($p < 0.05$)	17 (27)	48 (42)

Abbreviations: NS = not significant, IVC = inferior vena cava.

* Numbers in parentheses are percentages.

† These patients complained of abdominal venous distension as a symptom.

Liver histology

The liver biopsy specimens obtained during life were examined for: (i) centrizonal congestion, hemorrhage, and cell necrosis; and (ii) fibrosis, scarring, reversed lobulation, and the presence of regenerative nodules suggesting cirrhosis (Table 6).

While it was possible to assign most of the patients to either group (i) or group (ii), a significant degree of overlap was also observed (Table 6). As expected, congestion predominated in patients with acute BCS (71%), while those with chronic disease tended to have cirrhosis (67%). However, the presence of established cirrhosis in patients with acute BCS and purely congestive features in patients with chronic BCS remains to be explained. This clinicopathologic dissociation was documented earlier and emphasized by Parker (80). Representative histology is shown in Figures 1 and 2.

The diagnosis of hepatocellular carcinoma (HCC) was made during life in 5 patients in the chronic group; all had established cirrhosis as well. In all, 9 patients with HCC were recorded in this study, 4 of whom were positive for hepatitis B surface antigen (HBsAg). One patient each had a renal cell carcinoma and a metastatic anaplastic carcinoma.

Vascular anatomy

PTHV and IVCC were used to delineate the extent and location of hepatic venous outflow oc-

TABLE 6. Hepatic histopathology in acute and chronic Budd-Chiari syndrome

	Acute (%)	Chronic (%)	P
Centrizonal congestion & necrosis	71	29	<0.001
Mixed features; congestion & cirrhosis	18	4	<0.01
Established cirrhosis	11	67	<0.001

FIG. 1. Photomicrograph showing marked centrizonal congestion with sinusoidal dilatation (hematoxylin and eosin, orig. mag. × 220).

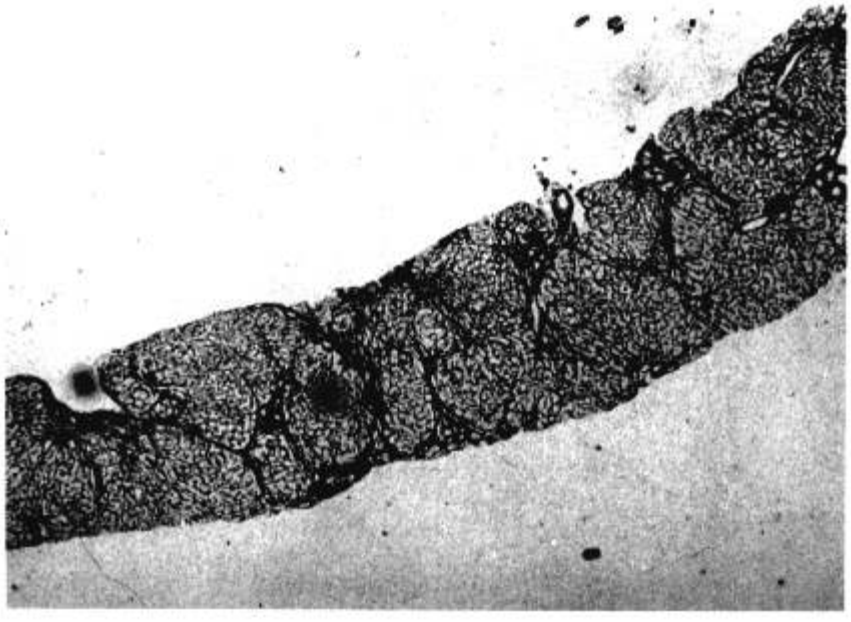

FIG. 2. Photomicrograph showing multiple small nodules surrounded by thin, fibrous septae. In contrast, other types of cirrhosis are marked by thick, fibrous septae (Gordon and Sweet stain, orig. mag. × 110).

clusion. Of the 119 patients who underwent both studies, 28 (24%) had pure HV block, 24 (20%) had pure IVC block, and 67 (56%) had both HV and IVC block (Table 7).

Radiologic features of HV block included ostial narrowing, partial obliteration, and complete nonvisualization. The formation of a characteristic "spider web" at attempted HV catheterization with contrast injection was seen in many patients, while intralobar, interlobar, and transhepatic collaterals (22) were observed in some. Patterns of IVC block included thin membranes or webs (MOVC), irregular filling defects suggesting thrombosis, segmental narrowing, or a complete nonvisualization of the retro- and suprahepatic IVC. Splenoportovenography was performed in a few patients with features of portal hypertension, revealing a hepatopetal flow of blood (21). The portal vein was not visualized in 2 patients. A few patients also underwent transhepatic assessment of hepatic vein pressure (83).

Patients with HV block and IVC block were dissimilar in several respects (Table 7). While the clinical features of acute BCS predominated in the former group, the latter was characterized by a long

TABLE 7. Clinical features of 119 patients who underwent both HV and IVC study*

	HV Block (N = 28)		IVC Block (N = 24)	Both Blocked (N = 67)	Total 119
Male:Female	1:1.5		3:1	1.5:1	1.3:1
Age at presentation (yrs)					
<18	5 (18)		4 (17)	6 (9)	15 (13)
19–49	20 (71)		19 (79)	58 (87)	97 (81)
>50	3 (11)		1 (4)	3 (4)	7 (60)
Duration of illness (mos.)					
<6	12 (43)	($p < 0.05$)	1 (4)	19 (28)	32 (27)
6–59	14 (50)		16 (67)	32 (48)	62 (52)
>60	2 (7)	(NS)	7 (29)	16 (24)	25 (21)
Abdominal pain	18 (64)	($p < 0.05$)	7 (29)	43 (64)	68 (57)
Distension	25 (89)	($p < 0.05$)	13 (54)	64 (95)	102 (86)
Jaundice	4 (14)	(NS)	1 (4)	17 (25)	22 (18)
Upper G.I. bleeding	7 (25)	(NS)	9 (37)	11 (16)	27 (23)
Distended veins†	10 (36)	($p < 0.05$)	17 (71)	31 (46)	68 (49)
Hepatic encephalopathy	4 (14)	(NS)	3 (12)	7 (10)	14 (12)
Hepatomegaly	24 (88)	(NS)	19 (79)	63 (94)	106 (89)
Splenomegaly	7 (25)	($p < 0.005$)	16 (67)	28 (42)	51 (42)
Edema	3 (11)	($p < 0.05$)	9 (37)	37 (56)	49 (41)
Serum bilirubin > 1 mg/dl	17 (61)	($p < 0.01$)	6 (25)	38 (58)	61 (51)
Serum albumin < 3 g/dl	14 (50)	(NS)	13 (54)	41 (62)	68 (37)
Prothrombin index < 70%	8 (29)	(NS)	2 (8)	13 (19)	23 (19)

Abbreviations: HV = hepatic vein, IVC = inferior vena cava, NS = not significant.
* Numbers in parentheses are percentages.
† These patients complained of abdominal venous distension as a symptom.

illness culminating in the development of portal hypertension (i.e., splenomegaly and distended veins). The group with both vessels blocked had features of both syndromes, yet the features were closer to those associated with HV block. At histology, features of centrizonal congestion and necrosis, or cirrhosis, were documented in 86% and 8% in patients with hepatic vein block, respectively; in 70% and 27% in those with both vessels blocked; and in 38% and 57% in patients with IVC block. Eleven of 28 patients (39%) with HV block had pregnancy-related BCS, and this etiologic factor was responsible for BCS in 10 of 67 patients (15%) with both vessels blocked. None of the patients with IVC block alone had pregnancy-related BCS.

Once US and CT became available, we compared the relative efficacy of these modalities against angiographic procedures in delineating the vascular lesions of BCS (unpublished observations). The study established the superiority of US over CT. Using angiographic findings as a "gold standard", ultrasound could correctly diagnose the site and type of block in 14 of 15 patients (93%), while computerized tomography was correct in only 5 (33%). The major abnormalities observed with US included nonvisualization of the hepaticocaval confluence, comma-shaped intrahepatic collaterals, abnormally thickened or irregular hepatic veins, and thrombus or membrane in the IVC. At present, with the availability of pulsed Doppler US, ultrasonography has become the first modality used to evaluate such patients. Computerized tomography is used only if US suggests the presence of a tumor. Facilities for magnetic resonance imaging (MRI) are not available.

Membranous obstruction of the IVC (MOVC)

Fifty-four patients had an angiographically demonstrated IVC web or membrane (Table 8). Ten (18.5%) of these had acute BCS while the rest had a chronic disorder. There was an overall male preponderance of 1.6:1, and no patient with MOVC presented before age 12. Although 5 patients had become symptomatic by that age, they did not present for investigation until later. Five additional patients developed symptoms between the ages of 12 and 18, but they also presented later—usually after 5 to 10 years of symptoms.

Clinical features of MOVC-related Budd-Chiari syndrome were similar to those observed overall, but splenomegaly was a more marked feature than in the other subgroups. Six patients with MOVC had a hepatocellular carcinoma (11%), a figure much lower than the reported incidence (72, 84, 91). Two of these 6 patients were HBsAg-positive.

Pregnancy-related BCS

Relationship with pregnancy and early puerperium was a striking feature encountered during this study (Table 9). Although most (68%) presented

TABLE 8. Membranous obstruction of the inferior vena cava (MOVC) in 54 patients with Budd-Chiari syndrome*

Number	Acute 10		Chronic 44
Male:Female	2.3:1		1.5:1
Age at presentation (yrs.)			
<12	0		0
12–18	1	(NS)	7
>18	9	(NS)	37
Abdominal pain	8 (80)	(NS)	18 (41)
Distension	9 (90)	(NS)	31 (70)
Jaundice	5 (50)	($p < 0.005$)	3 (7)
Distended veins†	0	($p < 0.001$)	27 (61)
Hepatic encephalopathy	1 (10)	(NS)	4 (9)
Upper G.I. bleeding	2 (20)	(NS)	11 (25)
Hepatomegaly	7 (70)	(NS)	39 (88)
Splenomegaly	5 (50)	(NS)	30 (68)
Ascites	9 (90)	(NS)	29 (59)
Edema	4 (40)	(NS)	15 (34)
Associated HV block	6 (60)	(NS)	20 (45)

Abbreviations: NS = not significant, HV = hepatic vein.
* Numbers in parentheses are percentages.
† These patients complained of abdominal venous distension as a symptom.

TABLE 9. Pregnancy- and puerperium-related Budd-Chiari syndrome in 38 patients*

Number	Acute 26		Chronic 12
Mean duration (mos.)	1.8		52.7
Abdominal pain	22 (88)	(NS)	12 (100)
Distension	21 (84)	(NS)	10 (85)
Jaundice	7 (28)	(NS)	1 (9)
Upper G.I. bleeding	3 (12)	(NS)	3 (26)
Distended veins†	1 (4)	($p < 0.001$)	10 (85)
Hepatic encephalopathy	7 (28)	(NS)	3 (26)
Hepatomegaly	19 (76)	(NS)	8 (67)
Splenomegaly	5 (20)	($p < 0.005$)	9 (75)
Ascites	24 (96)	(NS)	10 (85)
Edema	6 (24)	($p < 0.05$)	7 (59)
HV block	21/25	(NS)	10/12
IVC block	5/16	(NS)	6/12
Hospital deaths	18 (69)	($p < 0.01$)	2 (17)

For abbreviations, see earlier tables.
* Numbers in parentheses are percentages.
† These patients complained of abdominal venous distension as a symptom.

with acute symptoms, a significant number also had chronic BCS. This finding of chronic BCS has not, to our knowledge, been described previously (47, 61, 80, 101, 107, 109, 110). Pregnancy-related acute BCS was usually a fulminant disorder. These patients had a higher incidence of pain, gastrointestinal bleeding, and hepatic encephalopathy when compared to others with acute BCS due to non-pregnancy-related causes. They also had a significantly lower incidence of visible abdominal venous engorgement and splenomegaly. Eighteen of the twenty-six patients (69%) with acute BCS either died in hospital or left in extremis. Overall, pregnancy-related BCS was responsible for 20/36 (56%) of the deaths in the entire series of patients. Two patients with chronic BCS in this group received mesoatrial shunts. One died; the other is well more than 7 years later.

Budd-Chiari syndrome in childhood and adolescence

Thirty-eight patients (21%) developed BCS during childhood (onset before age 12) or adolescence (onset between ages 12 and 18) (Table 10). However, only 9 (5%) presented during childhood, 14 (8%) during adolescence, and 15 (8%) during early adulthood. There was an overall male preponderance of 3:2. Presentation of the acute form of BCS was distinctly uncommon. In the entire study population of 177 patients, the ratio of acute to chronic BCS was 5:9, but in childhood and adolescent BCS, it was 1:7. Hepatic histopathology revealed congestion in 27% and cirrhosis in 73%.

The proportion of patients with symptom duration of longer than 5 years at presentation was 66% in childhood BCS and 40% for the entire group. The younger patients also had a higher incidence of visibly dilated abdominal veins, and splenomegaly. In addition, these patients had a lower incidence of encephalopathy when compared to the older patients (0.3% vs 6.1%), possibly because younger patients had a larger number of rapidly opening collateral veins that allowed the early re-establishment of effective drainage for venous blood.

The incidence of MOVC in this group was not significantly higher than in the overall group. Three teenaged pregnancies led to the development of chronic BCS, and there were 2 MOVC-related hepatocellular carcinomas in this group. Two patients had shunt surgery but died.

Therapy for BCS

Fifteen, 10, and 2 patients, respectively, underwent meso-atrial or mesocaval shunt surgery, transcardiac "finger fracture" (membranotomy), and balloon membranotomy. There were 5 deaths in the early postoperative period in the shunt group (33%), 1 (10%) in the transcardiac membranotomy group, and 1 in the balloon membranotomy group.

Shunt surgery is expensive because the cost of materials is high. Hence, only a few patients could be operated upon. No patient has had thrombolytic therapy so far.

Discussion

Patients with Budd-Chiari syndrome can be divided into etiologic (Tables 4 and 11), anatomic

TABLE 10. Budd-Chiari syndrome in children and adolescents*

	Acute	Chronic Child†		Adolescents‡	
Duration of illness (months)	<6	6–59	>60	6–59	>60
Number	5	7	12	4	10
Male:Female	4:1	2:5	11:1	3:1	3:7
Age at presentation (yrs.)					
<12	4	4	1	0	0
12–18	1	3	6	4	0
19–30	0	0	5	0	9
Abdominal pain	4 (80)	2 (29)	6 (50)	2 (50)	6 (60)
Distension	5 (100)	6 (86)	8 (67)	1 (25)	6 (60)
Jaundice	3 (60)	1 (14)	2 (17)	0	1 (10)
Upper G.I. bleeding	0	1 (14)	3 (25)	1 (25)	3 (30)
Distended veins§	0	6 (86)	12 (100)	2 (50)	7 (70)
Hepatic encephalopathy	0	0	0	0	1 (10)
Hepatomegaly	5 (100)	6 (86)	11 (92)	2 (50)	10 (100)
Splenomegaly	1 (20)	4 (56)	12 (100)	2 (50)	5 (50)
Ascites	5 (100)	7 (100)	9 (75)	1 (25)	7 (70)
Edema	4 (80)	2 (29)	4 (33)	0	6 (60)
HV block	3/3	5/5	5/8	2/2	5/6
IVC block	5/5	5/7	12/12	2/2	8/9
Etiology					
Membranous obstruction of the inferior vena cava	0	0	5	2	5
Pregnancy	0	0	0	0	3
Hepatoma	0	0	2	0	0

For abbreviations see earlier tables.
* Numbers in parentheses are percentages.
† Disease onset <12 years of age.
‡ Disease onset between 12 and 18 years of age.
§ These patients complained of abdominal venous distension as a symptom.

TABLE 11. Etiology of Budd-Chiari syndrome: Comparative data

Reference	80	70	61	72	109	Present study
Country	*	*	France	Japan*	China	India
Patients (no.)	164	243	38	100	100	177
Etiology (%)						
Tumors	13	9	5	6	1	6
Infections	5	10	0	12	0	3
Pregnancy/ OC†	3	20	13	0	1	21
Blood dyscrasia	12	18	47	0	2	4
MOVC	0	‡	5	64	37	29
Miscellaneous	3	13	2	11	7	4
Idiopathic	64	30	26	7	51	35

For abbrevation, see previous tables.
* Collective review.
† Use of oral contraceptives.
‡ MOVC excluded.

(Table 7), histologic (Table 6), and clinical groups (Table 5). Such compartmentalization has clarified some of the complex interactions between the major determinants of the outcome of this condition, both with and without therapeutic intervention (55).

Etiology, pathogenesis, and clinical presentation

Although the etiology and pathogenesis of BCS are not yet completely understood, BCS is a primarily vascular disorder, and the presence of a hypercoagulable state thus appears inevitable. Although coagulopathies have been reported in a few patients (6, 7, 13, 55, 69), most patients had no demonstrable coagulopathy at presentation (35), suggesting that some of these mechanisms may be intermittent or even transient in nature (82).

Several conditions associated with hypercoagulability have been implicated in the causation of BCS (Table 12). The natural history of many of these conditions could also be reflected in the clinical manifestations of BCS. Conditions with a "one-time" hypercoagulability, such as pregnancy, infections, tumors, trauma, and vascular injury (Table

TABLE 12. Etiology of Budd-Chiari syndrome: Conditions documented in the literature

Condition	References
Tumors	
Adrenal carcinoma	19, 80
Atrial tumor	58
Bronchogenic carcinoma	80
Hepatoma	58, 60, 72, 91, 109
Leukemia	80, 86
Leiomyoma	29, 54
Sarcoma of IVC	65
Lymphoma	65
Pancreatic carcinoma	80
Renal cell carcinoma	80, 94
Stomach carcinoma	70
Wilms tumor	89
Infections	
Amebic liver abscess	1, 47
Aspergillosis	80
Filariasis	106
Hydatid cysts	1
Pyemic liver abscess	1
Pelvic cellulitis	80
Schistosomiasis	80
Syphilis	70, 72
Tuberculosis	72, 80, 105
Zygomycete	102
Pregnancy	47, 61, 70, 80, 101, 107, 110
Oral contraception	53, 58, 70, 93, 101
Blood dyscrasias	
Antithrombin III deficiency	55
Lupus anticoagulant syndrome	7
Myeloproliferative syndrome	3, 58, 70, 80, 99
Paroxysmal nocturnal hemoglobinuria	6, 70, 80, 100
Protein C deficiency	13
Sickle cell disease	69, 80
MOVC	23, 25, 31, 41, 51, 72, 76, 78, 91, 103, 109
Miscellaneous	
Alpha-1-antitrypsin deficiency	58
Behçet's disease	9, 109, 111
Crohn's disease	56
Coarctation of inferior vena cava	104
Congenital anomalies	5, 72, 92, 109
Granulomatous hepatic angiitis	115
Hypereosinophilic syndrome	58
Mixed connective tissue disease	24
Nodular regenerative hyperplasia of liver	20
Sarcoidosis	58
Sjögren syndrome	62
Systemic lupus erythematosus	28
Total parental nutrition	64
Trauma	58, 70, 80, 85
Ulcerative colitis	15, 80
Ventriculoatrial shunt	79

12), are most likely to produce the acute form of BCS (Table 4; refs. 58, 70, 80). Diseases with a prolonged course (Table 12) are more likely to lead to an indolent form of BCS, although an acute presentation cannot be ruled out completely.

The discrepancy between the clinical course and the histology, observed both previously (80) and in our study, can be explained: The presence of cirrhosis and reversed lobulation in patients with acute BCS must be due to earlier asymptomatic, or only mildly troublesome, episodes of hepatic venous outflow obstruction. Similarly, the finding of congestion alone in liver biopsies of patients with chronic BCS can be attributed to sampling from a recently affected area of the liver. The clinical presentation may result from either a large, sudden blockage (acute BCS) or the development of portal hypertension through an insidious gradual outflow obstruction (chronic BCS). Thus, the natural history of BCS probably reflects the rate of progress of the primary underlying thrombotic disease process, and the balance between it and the efficiency of the restorative thrombolytic process. While the natural thrombolytic processes may facilitate the recanalization of a blocked outflow channel, serial biopsy studies have shown the injury's fibrogenic potential to be unaffected (40). Consequent hemodynamic alterations initiate and regulate the subsequent opening of collaterals in an attempt to decompress the liver parenchyma. These processes can explain the wide spectrum of presentation in BCS (53, 77).

A recent French study (99) elegantly demonstrated an occult myeloproliferative syndrome in almost 80% of patients in a group believed to be suffering from idiopathic BCS. Their study was based on the spontaneous generation of colony-forming units from the bone marrow at culture. However, the failure of these patients to progress to an overt myeloproliferative syndrome over an extended period of observation does not support this contention (99), a finding underscored by a German study of 500 patients with established myeloproliferative syndromes, in which only 6 (1%) developed BCS (3). Although occult myeloproliferative disorders are a conceptually attractive hypothesis to explain "idiopathic" chronic or acute BCS, we believe that their etiologic and pathogenetic role in BCS must be more widely established and a detailed study of their natural history undertaken to clarify their exact nature.

Membranous obstruction of the IVC (MOVC) is a frequent finding in patients with chronic BCS in Asia (41, 48, 72, 75, 114), India (25, 27, 47), and South Africa (91). The origin and nature of this membrane are disputed (23, 41, 76). Protagonists of the "acquired origin" theory have demonstrated the appearance of a membrane from what was a thrombus (97). They have also emphasized the wide variability in the anatomic location of the membrane (16, 23), its variable thickness (4), and its late presentation in most patients (during the fourth decade of life) as evidence to refute the "congenital anomaly" theory (76). Histologic evi-

dence also suggests that the membrane may be an acquired lesion (76). Nevertheless, MOVC has been seen in young children in our own and other studies (42, 104). We believe that although most of these membranes may be acquired, the complex developmental anatomy of the region makes it a perfect site for a congenital defect (104).

Mitchell et al (70) excluded MOVC from their review of the etiology of 243 patients with BCS. However, we believe that since MOVC is an anatomically demonstrable anomaly observed worldwide (72, 84, 91, 109), offering itself to relatively simple surgical (4, 48, 84, 87) or other interventional (30, 95, 113) amelioration, and having a documented though debatable association with hepatocellular carcinoma (72, 84, 91), it should be maintained as a separate subgroup of BCS pending a final decision on its origin.

Studies from different parts of the world (Table 11) have shown that the predominant cause of BCS differs by locale:, being MOVC in Asia and blood dyscrasias in Western countries.

Pregnancy and puerperium-related BCS

Pregnancy-related BCS is common in India (1, 47) and has been documented since the turn of the century (80). We believe that several factors contribute to this common occurrence in our country. First, pregnancy is a physiological hypercoagulable state with increased serum clotting factors of predominantly hepatic origin (70). These factors enter the circulation through the hepatic veins, where their concentration would be the highest. Second, in rural India, prolonged rest postpartum is followed by late and slow mobilization after childbirth, up to 40 days of confinement, and fluid restriction (believed to enhance the quality of the milk). This combination of increased clotting factors and dehydration prepares an ideal setting for thrombotic complications. It may also account for the increased incidence of postpartum cerebral cortical venous thrombosis observed in our area.

The most common presentation in this group of patients was that of fulminant BCS (47), but we also observed a previously unreported chronic form, probably stemming from a partial block that allowed time for portal hypertension to develop.

Membranous obstruction of the inferior vena cava (MOVC)

Membranous obstruction of the inferior vena cava (Table 13) has been described predominantly in China (109), Japan (41, 48, 72, 75, 114), India (1, 25, 27, 47), and South Africa (42, 91), but a few patients have also been recorded in the United States (84), Europe (61), and other developed nations. This abnormality has usually been associated with chronic BCS, but 10 of our patients presented with acute BCS (Table 8). We believe that this acute presentation was due to the development of a fresh thrombus on a pre-existing IVC membrane.

The clinical manifestations in our patients were similar to those recorded in the literature except that our patients did not have a high incidence of hepatocellular carcinoma (HCC). The earlier Japanese literature (41, 72) and a report from South Africa (91) noted a very high incidence of malignancy, which was also observed in a few patients in the United States (84). Both of our childhood patients with cancer had a coexisting membrane, but the overall incidence of HCC was low. Hepatoma was noted in only 1 of 37 Chinese patients (3%) (109) and in only 9 of 155 recently observed Japanese patients with MOVC (75). Our findings show a similar trend. Okuda et al (75) emphasized that whereas BCS was found in 20% of hepatomas seen at autopsy in South Africa, only 2 of 2,054 Japanese cases with HCC had coexisting BCS. These observations, and the lack of information regarding the prevalence of hepatitis virus infections in earlier patients (91), makes it difficult to attribute malignancy to MOVC alone—or to the degree previously reported (41, 72, 91).

Vascular anatomy of Budd-Chiari syndrome

Two distinct groups can be identified based on the vascular anatomy of BCS: patients with HV block associated with symptoms and signs of acute BCS and caused by conditions promoting hypercoagulability (e.g., blood dyscrasias of pregnancy); and patients with IVC block dominated by a clinical picture of chronic BCS and caused by MOVC. However, most of our patients (56%) had both HV and IVC block. The dominant lesion determining the severity of BCS was hepatic vein block, because this block had the maximal effect on liver cell function and architecture. It was thus not surprising that the clinical manifestations of patients with both blocked vessels closely resembled those of patients with pure HV block rather than those with pure IVC block (57, 70). Defining the vascular anatomy is a crucial step in deciding upon the surgical procedure to be undertaken (50). There can be no debate regarding the necessity to establish the patency of the IVC before planning a surgical procedure in patients with BCS (16, 40, 50), and angiography remains the "gold standard" against which all other modalities are assessed.

Of the newer modalities, US has emerged as the most reliable (8, 11, 12, 36, 43, 59, 67, 71, 74). Its relatively low cost and ease of repetition are added

TABLE 13. MOVC: Comparative analysis from 5 geographical areas

Reference	72	91	84	109	Present study
Country	Japan	S. Africa	U.S.A.	China	India
MOVC/Total	64/107	101/101	8/8	37/100	54/177
Male:Female	1:1	3:1	3:1	3:2	5:3
Age at diagnosis (yrs.)	27	—	33	33	30
Duration of symptoms (mos.)					
<6	—	—	—	—	19
6–59	—	—	—	—	41
60–119	—	—	—	—	17
>120	—	—	—	—	23
Clinical findings (%)					
Abdominal pain	10	—	50	—	48
Distension	—	—	—	—	75
Jaundice	12	—	—	—	15
Upper G.I. bleeding	11	—	13	22	24
Hepatic encephalopathy	15	—	—	8	9
Hepatomegaly	37	92	100	99	87
Splenomegaly	32	40	50		65
Engorged veins	70	71	63	100	81
Ascites	77	62	37	76	65
Edema	54	60	87	—	35
Hepatoma	36	49	25	1	11

For abbreviation, see previous tables.

assets. The important abnormalities in BCS detectable by US are nonvisualization of hepatic veins (12, 58, 59, 67, 71), areas of stenosis, ostial obstruction (presence of membrane), irregularity suggestive of thrombosis, prominence of collateral veins, and proximal dilatation (71). Doppler demonstration of altered or absent hepatic vein flow has added significantly to the diagnostic accuracy (12, 36, 43, 71, 74). Computerized tomography is helpful only in selected circumstances but MRI may become the most accurate investigation (33, 34, 71, 96). The high cost of the latter would, however, be a disadvantage. A dynamic CT examination is more informative than plain or enhanced (8, 61, 71) scans alone.

Natural history

Our study has confirmed the presence of 2 basic types of presentation in Budd-Chiari syndrome. The acute form, presenting with hepatocellular necrosis or failure, was characterized by pain, ascites, jaundice, encephalopathy, tender hepatomegaly, and the presence of hepatic vein block either in isolation or with IVC block (57, 70, 80). This acute syndrome was most frequently attributed to pregnancy and other hypercoagulable states.

One-third of our patients presented with the acute form of BCS. Histologically, the dominant lesions were centrizonal congestion and necrosis. The mortality was high in those with extensive block and a paucity of collateral veins, although some patients recovered from the acute episode and presented several years later with features of chronic BCS and cirrhosis.

The chronic syndrome, which occurred in two-thirds of our patients, was more varied in its presentation than the acute, but was dominated by features and complications of portal hypertension (41, 72). Although histologically, most of these patients were shown to have cirrhosis, their liver functions were better preserved than in patients with postnecrotic or alcoholic cirrhosis. Most patients in this group timed their complaints to an acute attack, which gradually abated, to be followed by several months of apparently good health before the insidious development of ascites and visible veins over the abdomen. These problems frequently brought patients to hospital more than 5 years (and often more than 10 years) after the initial event. Moreover, if the initiating and subsequent thrombotic events were localized and insidious, patients were sometimes completely symptom-free until they presented with signs and symptoms of cirrhosis.

Budd-Chiari syndrome in children

Acute BCS is uncommon in children, although the chronic presentation is proportionally more frequent than in adults. This may reflect a more effective establishment of collateral channels or a more vigorous recanalization process at a younger age. The clinical manifestations were similar to those observed in adults with BCS.

Only 2 major reports of BCS in childhood are available (35, 42). In comparing our data to these reports (Table 14), we found that the diverse backgrounds of other patients obscured the etiology of childhood BCS. The French study (35) included

TABLE 14. BCS in children: Comparative analysis

Reference	35	42	Present study 24	
Number of patients	22*	17†	Acute (5)	Chronic (19)
Male:Female	6.5	4.6:1	4:1	2:1
Etiology (%)				
MOVC	0	100	0	26
Hepatoma	0	0	0	11
HVOD	46	0	0	0‡
Clinical features (%)				
Abdominal pain	19	—	80	42
Distension	46	36	100	74
Jaundice	—	—	60	16
Upper G.I. bleeding	—	—	0	21
Distended veins§	73	64	0	95
Hepatic encephalopathy	0	0	0	0
Hepatomegaly	100	100	100	89
Splenomegaly	54	50	20	84
Ascites	63	—	100	84
Edema	—	—	80	32
HV block (no./total)	19/22	—	3/3	10/13
IVC block (no./total)	8/20	17/17	5/5	17/19

Abbreviation HVOD = hepatic veno-occlusive disease. For other abbreviations, see earlier tables.
* 3 with acute, 19 with chronic disease.
† All with chronic disease.
‡ HVOD excluded.
§ These patients complained of abdominal venous distension as a symptom.

several patients from North Africa with HVOD. Many of them had been exposed to pyrrolizidine alkaloids, and we excluded this group from our study. The study from South Africa (42) reported a very high incidence of MOVC, which we have not observed in our group of patients. Although the presence of so many symptomatic children with MOVC suggests a congenital basis for membrane formation, this hypothesis clearly needs further study.

Treatment

Several surgical and other interventional techniques have been used to treat BCS (Table 15). Shunt surgery attempts to provide adequate hepatic decompression using the portal venous system as an improvised outflow tract (40, 46, 50, 70, 77). Depending upon the accessibility of various vessels and the skill, experience, and ingenuity of the surgeon, several procedures can be used (Table 15). Survival and clinical improvement have followed all these procedures (40, 50, 70, 78, 107, 109, 116, 117), but most do not attempt to correct the underlying vascular defect. The Senning procedure (dorsal resection with anastomosis) (73, 90, 108) is one of the newer operations attempting to do so (112). This procedure consists of a direct hepatoatrial anastomosis created after resection of the portion of the liver containing the terminal portions of the major hepatic veins; this procedure allows hepatic venous blood to enter the heart. Hepatic transplantation has also been performed successfully (38, 44, 88).

Current trends suggest that surgical correction of BCS should be performed as soon as possible after diagnosis (50, 70, 77). The choice of surgical procedure is determined by the site and extent of the blockage (Table 16). The most widely used shunts are the mesocaval and portocaval for HV block and the mesoatrial for both HV and IVC block. The long-term results of liver transplantation are comparable to those of shunt surgery. We believe that transplantation is the preferred procedure in patients with fulminant hepatic failure due to acute BCS and in those with advanced decompensated cirrhosis.

The rapid developments in interventional techniques offer hope to several other groups of patients. In acute BCS, local infusions of thrombolytic substances at the site of block followed by stent insertion, if required, may be curative. In patients with chronic BCS, the creation of an intrahepatic shunt between the hepatic veins and a branch of the portal vein may provide for adequate decompression of the liver.

The patency of the IVC is central to the selection of a therapeutic approach. If it is patent and a gradient of > 20 mmHg exists between the portal system and the IVC, then a mesocaval or portocaval procedure would decompress the liver. If a short segment of the IVC is blocked, intervention with a balloon is justified, provided the major hepatic veins are patent (Table 16). If both the IVC and hepatic

TABLE 15. Therapeutic options in Budd-Chiari syndrome: Methods described in literature

Options	Reference
Medical	
Diruetics	
Thrombolysis: Streptokinase	4, 57, 58
Urokinase	37
Anticoagulants	53
Surgical procedures	
Shunt Procedures	
Portocaval	10, 77, 78, 81, 110, 116
Mesocaval	16, 49, 50, 109, 110, 116
Mesoatrial	16, 17, 40, 49, 50, 109, 110
Portoatrial	58, 107
Cavoatrial	78, 107, 109
Meso-pulmonary vein	40
Splenoatrial	40, 109
Mesoiliac	116
Mesocavoatrial	109
Two-stage shunts	46, 110
Non-shunt procedures	
Transcardiac membranotomy	41, 48, 84, 87, 114
Excision of membrane	41
Bypass graft	41, 104
Hepatopneumopexy	41
Splenopneumopexy	2, 41
Senning operation	73, 90, 108
Dorsal resection	112
LeVeen shunt	65
Liver transplantation	18, 39, 44, 88
Interventional radiology	
Angioplasty	30, 95
Membranotomy	103
Angioplasty of post-shunt ostial stenosis	46
Laser angioplasty	113
Intravascular saw	113
Stents (expandable, retaining, retrievable)	113

veins are blocked but the IVC block closes off only a short segment, balloon dilatation and a mesocaval or portocaval shunt would be acceptable. If those procedures fail, a mesoatrial shunt would be the procedure of choice.

Our experience with these surgical procedures has been limited, because of their high cost: The mesoatrial shunt alone costs more than the average yearly income of our patients. The need to develop a low-cost procedure is thus pressing.

Summary

Budd-Chiari syndrome (BCS) may not be as uncommon as was once believed. Our study has substantiated the existence of 2 major clinical forms. The acute syndrome is invariably associated with extensive blockage of the major hepatic veins, resulting in congestive liver cell necrosis. In a small, but significant, number of patients the inferior vena cava (IVC) is also occluded. The important etiologic factors are related to hypercoagulability of blood. Immediate placement of a shunt improves survival. The chronic syndrome is characterized by portal hypertension and is associated with a variable abnormal vascular anatomy. The causes of the chronic syndrome are not clear, but a substantial number of cases are related to the presence of an IVC membrane. Shunt surgery is effective but procedures aimed at the primary pathology are likely to be even more so.

The natural history of BCS should be viewed over a long period of time. The very long survival of several patients urges a more cautious approach to surgical remedies. Budd-Chiari syndrome probably represents a spectrum of disease caused primarily by a hypercoagulable state and having a varied presentation depending on the balance between rate of formation and the extent of the thrombosis and the body's own rate of thrombolysis and recanalization. The extent and efficacy of the individual's collateral circulation and the rate of development of liver fibrosis are other determinants. It is thus possible to view BCS as a continuum of a single pathogenetic spectrum.

Pregnancy-related BCS in India probably has strong social determinants, and is usually acute and fulminant. We have, however, documented a chronic form not described earlier. Children usually do not have acute BCS, but chronic BCS in children and adolescents is similar to that in adults. Membranous obstruction of the inferior vena cava (MOVC) is common and was found even at a young age. The association of MOVC with hepatocellular carcinoma, however, did not appear to be as clear as was previously believed.

TABLE 16. Suggested treatment plan in Budd-Chiari syndrome

Region	Extent of obstruction	
	Short segment	Long segment
Hepatic vein	Radiological intervention Portocaval or mesocaval shunt	Portocaval or mesocaval shunt
Inferior vena cava	Radiological intervention Transcardiac membranotomy Cavoatrial shunt	Mesoatrial shunt
Both	Radiological intervention Portocaval or mesocaval shunt with cavoatrial shunt	Mesoatrial shunt

There has been a wide geographical variability in the causes and manifestations of BCS. Our study has clearly shown that—Kipling's categorical statement to the contrary—East and West do meet in India, in the Budd-Chiari syndrome.

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