Current Management of the Budd–Chiari Syndrome

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Twenty-six patients with the Budd–Chiari syndrome were treated surgically at the Johns Hopkins Hospital. Twenty-one of the patients were female and five were male, with a median age at diagnosis of 37 years. Nine patients had polycythemia vera, 6 were receiving estrogen therapy, 5 had a previous hepatitis A or B infection, and 4 had cirrhosis. There was one case each of hepatic malignancy, paroxysmal nocturnal hemoglobinuria, and idiopathic thrombocytopenic purpura. In five cases no etiologic factors or associated disorders were identified. Ascites was the most common presenting feature in this group of patients. Hepatic function at the time of diagnosis, as measured by standard serum chemistries, was only minimally abnormal. The diagnosis of the Budd–Chiari syndrome was confirmed in all 26 patients by hepatic vein catheterization. Inferior vena cavography was also performed and revealed caval occlusion in 4 patients, significant caval obstruction in 13 patients, and a normal vena cava in 9 patients. Interpretation of the vena cavogram was helpful in selecting the appropriate surgical procedure for each patient. Twenty-three of the twenty-six patients underwent percutaneous liver biopsy before operation, with no morbidity or mortality. Four patients had well-established cirrhosis noted on biopsy. Thirty mesenteric-systemic venous shunts were performed on the 26 patients. In 11 patients a mesocaval shunt was performed and in one instance conversion to a mesoatrial shunt was required as a second procedure. In 15 patients a mesoatrial shunt was performed as the initial procedure. Graft thrombosis occurring in 2 of these 15 patients prompted one revision in 1 patient and 2 revisions in the second patient. After mesenteric-systemic venous shunt, eight of the patients (31%) died before discharge from the hospital. The remaining 18 patients in this series were discharged from the hospital alive and well with patent shunts. Patients were followed for a median of 43 months (range, 9 months to 13 years). Five late deaths occurred between 5 and 84 months after the operation. Three- and five-year actuarial survival rates were 65% and 59%, respectively.

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THE BUDD–CHIARI syndrome is a rare, often fatal illness resulting from hepatic venous outflow occlusion. The obstruction may be due to thrombosis of the major hepatic veins or suprahepatic vena cava or, as is more common in the Orient, membranous obstruction of the suprahepatic inferior vena cava.[1] Patients with hepatic venous congestion secondary to the Budd–Chiari syndrome present with manifestations of portal hypertension (ascites and/or bleeding esophageal varices), which may develop either acutely or chronically. A number of surgical procedures have been suggested to decompress the congested liver by converting the portal vein into an outflow tract.[2–8] Portacaval, mesocaval, or mesoatrial shunts have all been used successfully. Orthotopic liver transplantation is now an option in managing the Budd–Chiari syndrome and selected patients can expect the most favorable outcome with this form of therapy.[9,10] We report here experience with 26 patients with the Budd–Chiari syndrome who were treated surgically at the Johns Hopkins Hospital during a 15-year period. Based on this experience we have developed a treatment regimen that should result in successful management of most patients with this otherwise fatal syndrome.

Clinical Material

Between 1973 and 1988, 26 patients with the Budd–Chiari syndrome were treated surgically at the Johns Hopkins Hospital. Twenty-one were women and five were men (Table 1). The median age at the time of diagnosis was 37 years (range, 14 to 68 years) and the median duration of symptoms prior to evaluation at the Johns Hopkins Hospital was between five and six months (range, 1 week to 5 years). Twenty-four of the 26 patients were found to have ascites at the time of diagnosis and in most cases the ascites developed over a period of weeks or

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TABLE 1. *Epidemiologic Factors in 26 Patients with Budd-Chiari Syndrome*

Factor	Mesocaval Shunt	Mesoatrial Shunt	Both
Age	32 years	40 years	37 years
Sex	9F/2M	12F/3M	21F/5M
Race	9W/2B	12W/3B	21W/5B
Symptom duration at diagnosis	6 months	5 months	5.5 months

TABLE 3. *Results of Preoperative Inferior Vena Cavography Among 26 Patients with the Budd-Chiari Syndrome*

Result	Mesoatrial Shunt	Mesocaval Shunt
Caval occlusion	4/15 (27%)	0
Significant caval obstruction (compression >75% or gradient >20 mmHg)	11/15 (73%)	2/11 (18%)
No significant caval obstruction	0	9/11 (82%)

months. Abdominal pain or discomfort, always mild, was present in 42% (11 of 26) of our patients. Jaundice was clinically evident in 27% (7 of 26) and gastrointestinal bleeding was documented in five patients (19%).

Of the etiologic factors known to be associated with the Budd–Chiari syndrome, polycythemia vera (35% or 9 patients) and estrogen use (23% or 6 patients) were most commonly recognized. Five of the twenty-six patients (19%) had previously documented episodes of infectious hepatitis and four patients (15%) were known to have cirrhosis at the time of diagnosis. Paroxysmal nocturnal hemoglobinuria, idiopathic thrombocytopenic purpura, and hepatic malignancy were each present in one patient. Five patients (19%) had no identifiable predisposing or associated factors for the Budd–Chiari syndrome and were categorized as idiopathic (Table 2).

At the time of admission the median serum bilirubin was 1.4 mg/dL (range, 0.6 to 25.0 mg/dL), median SGOT was 39 IU/L (range, 11 to 9480 IU/L), median SGPT was 22 IU/L (range, 14 to 6710 IU/L), and median alkaline phosphatase was 136 IU/L (range, 44 to 326 IU/L). Mild elevation of the prothrombin time was noted, with a mean value of 13.8 seconds (range, 11.5 to 19.7 seconds).

All patients were evaluated by catheterization and imaging of the hepatic veins. The diagnosis of the Budd–Chiari syndrome was confirmed by demonstrating completely occluded hepatic veins or recanalized veins with the typical 'spider web' appearance. The inferior vena cava was also evaluated angiographically, the results of which are shown in Table 3. Total caval occlusion was demonstrated in four patients. Significant caval obstruction, as demonstrated by more than 75% reduction of luminal diameter by extrinsic compression or intracaval thrombus, or a pressure gradient across the compressed cava exceeding 20 mm of mercury, was present in 13 patients. No significant caval obstruction was observed in nine patients.

TABLE 2. *Etiologic Factors in 26 Patients with the Budd-Chiari Syndrome*

Factor	Mesocaval Shunt	Mesoatrial Shunt	Both
Polycythemia	4 (36%)	5 (33%)	9 (35%)
Estrogen use	3 (27%)	3 (20%)	6 (23%)
Previous hepatitis	2 (18%)	3 (20%)	5 (19%)
Cirrhosis	1 (9%)	3 (20%)	4 (15%)
Hepatic malignancy	0	1 (7%)	1 (4%)
Paroxysmal nocturnal hemoglobinuria	1 (9%)	0	1 (4%)
ITP	0	1 (7%)	1 (4%)
Idiopathic	2 (18%)	3 (20%)	5 (19%)

ITP, idiopathic thrombocytopenic purpura

Percutaneous liver biopsies were performed before operation on 23 of the 26 patients. Despite the presence of ascites and a mild coagulopathy, there was no morbidity or mortality from these procedures. The histopathologic picture most often associated with the Budd–Chiari syndrome was centrilobular congestion. The spectrum of this pathology ranged from sinusoidal dilatation with variable amounts of red cell extravasation to hepatocellular atrophy and finally cell drop-out in the central vein regions. Nine patients had fibrotic changes noted on biopsy and four had cirrhosis. Two of the four patients with cirrhosis were thought to have alcoholic liver disease and two had postnecrotic cirrhosis (one hepatitis B and one non-A non-B hepatitis).

Operative Management

Thirty portosystemic shunt procedures were performed on the 26 patients included in this study. As the initial procedure, 11 patients underwent a mesocaval shunt and 15 underwent a mesoatrial shunt. Mesocaval shunts were performed using a 16-mm or 18-mm knitted Dacron graft in a 'C-type' configuration as described previously.[11] The first seven mesoatrial shunts used a 16-mm woven Dacron graft. The prosthesis was anastomosed first to the superior mesenteric vein in end-to-side fashion and then passed through the transverse mesocolon into the lesser sac. The graft then passed anteriorly through the greater omentum, and anterior to the stomach and left lobe of the liver, into the anterior mediastinum, and exited into the right chest. The atrial anastomosis was performed end-to-side through an anterior right thoracotomy. Concern that graft compression at the level of the sternum might result in reduced long-term patency was realized when reoperation for focal high-grade stenosis was required in a young woman 10 months after mesoatrial shunt. Since that time

and for the remaining eight patients, a modified 16-mm woven Dacron or ring-reinforced Gortex prosthesis, externally supported with an 8-cm silicone rubber cuff has been used.[12] The silicone cuff is positioned beneath the sternum as the graft traverses the diaphragm into the anterior mediastinum.

The decision as to which shunt to select was largely based on radiographic information. All four patients demonstrating total inferior vena cava occlusion were managed with a mesoatrial shunt. The nine patients without evidence of caval obstruction were treated with mesocaval shunts. If significant but not total vena caval obstruction was noted, as was the case in 13 patients, a mesoatrial shunt was the initial procedure for 11 patients and a mesocaval shunt was performed in 2 patients. One of these two patients who underwent a mesocaval shunt, despite a narrowed intrahepatic vena cava and caval gradient of 16 mmHg, required conversion to a mesoatrial shunt 2 weeks later. Two patients who underwent mesoatrial shunt as the initial surgical procedure required shunt revision at 6 months and 11 months after operation. In one instance a second revision was performed 5 years after operation.

Clinical Course

Eight of the twenty-six patients (31%) died before discharge from the hospital. The four patients who died within 30 days of their surgery were among the earliest treated at Johns Hopkins (before 1984). The first patient in the Johns Hopkins series was moribund at the time her mesoatrial shunt was performed and never awakened from her encephalopathic coma. The second death occurred in a woman who at autopsy was found to have metastatic bronchogenic carcinoma. The remaining two early perioperative deaths occurred in debilitated individuals with systemic fungal or bacterial sepsis before operation. Of the four deaths that occurred after the first 30 postoperative days, but before discharge, three were in women older than 64 years. The remaining patient, who was noted before operation to have significant vena caval compression, thrombosed her mesocaval shunt 2 weeks after operation, underwent revision to a mesoatrial shunt, and died 1 month later. Autopsy consent was obtained for five of the eight hospital deaths. In all five patients the portosystemic graft was determined to be patent at the time of death. Of the seven patients who have undergone portosystemic decompression for the Budd–Chiari syndrome since 1986, six were discharged from the hospital alive and well with patent shunts.

TABLE 4. *Follow-up of 10 Patients with the Budd–Chiari Syndrome Discharged Alive and Well After Mesoatrial Shunt*

Patient	Status	Months After Operation	Cause of Death
1	A & W	80	—
2	Died	75	Liver failure
3	Died	60	Bleeding duodenal ulcer
4	A & W	58	—
5	Died	40	Liver failure
6	A & W	36	—
7	A & W	29	—
8	Died	5	Liver failure
9	A & W	3	—
10	A & W	2	—

A & W, alive and well.
—, still alive at time of publication.

TABLE 5. *Follow-up of 8 Patients with the Budd–Chiari Syndrome Discharged Alive and Well After Mesocaval Shunt*

Patient	Status	Months After Operation	Cause of Death
1	A & W	150	—
2	A & W	132	—
3	Died	84	Unknown
4	A & W	46	—
5	A & W	37	—
6	A & W	24	—
7	A & W	6	—
8	A & W	2	—

—, still alive at time of publication.

Eighteen of the twenty-six patients were discharged alive and well with patent shunts. This included 8 of the 11 patients (73%) who underwent a mesocaval shunt and 10 of the 15 patients (66%) who underwent a mesoatrial shunt. Shunt patency was documented by angiography or more recently by computed tomography.[13] Median follow-up for the 18 survivors was 43 months (range, 9 months to 13 years). There were five late deaths, four in the mesoatrial group and one in the mesocaval group. Liver failure was responsible for three of these deaths, a bleeding duodenal ulcer for one, and in one case the cause of death was unknown. Two of the five late deaths occurred in patients with cirrhosis or evidence of pre-existing infectious hepatitis before their shunt procedure (Tables 4 and 5). Actuarial survival statistics for all Budd–Chiari patients treated surgically at The Johns Hopkins Hospital stratified by type of shunt performed is shown in Figure 1. The 3- and 5-year survival rates for all patients in the study were 65% and 59%, respectively. Patients who underwent a mesoatrial shunt had 3- and 5-year survival rates of 60% and 49%, respectively. The 5-year actuarial survival rate for the mesocaval group was 73%, with no deaths from time of discharge to the 5-year mark. One patient treated with a mesocaval shunt died of unknown causes 80 months after his surgery.

Over the past 4 years 10 patients with the Budd–Chiari syndrome have undergone mesoatrial (5 patients) or mesocaval (5 patients) shunts. Eight of these ten patients are

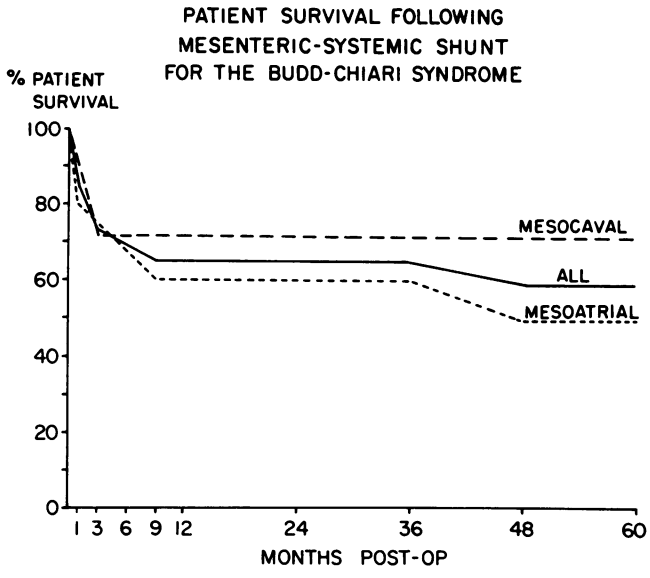


FIG. 1. Actuarial survival for patients with the Budd–Chiari syndrome treated by either mesocaval or mesoatrial shunt.

alive and well with patent shunts. Mean follow-up for this group of recently treated patients is 27 months (range, 9 to 49 months).

Recurrent ascites occurred in 6 of the 18 long-term survivors. Graft thrombosis was documented in four of these patients (three mesoatrial and one mesocaval). In the face of graft occlusion the ascites were managed by LeVeen shunt (two patients), redo mesoatrial shunt (one patient), or retrohepatic cavoplasty (one patient).[14] Two patients who underwent mesocaval shunt developed shunt stenosis at the venocaval anastomosis. They were successfully treated by transluminal angioplasty with resolution of the recurrent ascites. One patient with established cirrhosis at the time of mesoatrial shunt subsequently developed a hepatoma and underwent a successful orthotopic liver transplant.[15]

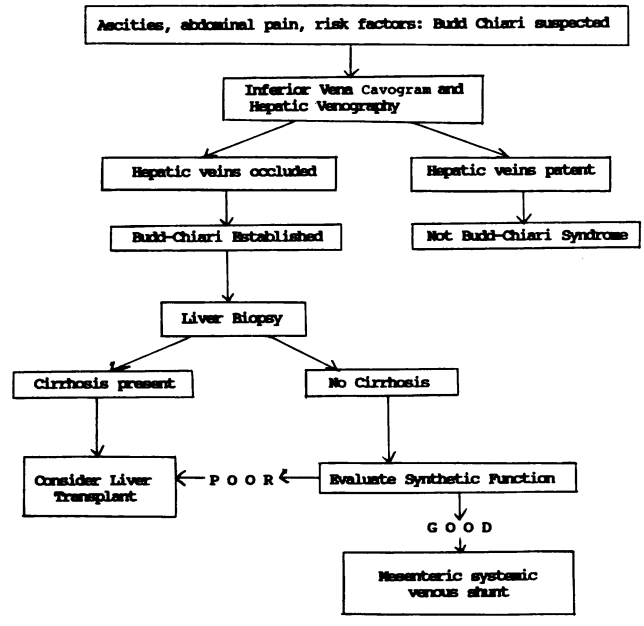
Discussion

Untreated hepatic venous thrombosis results in progressive liver failure and ultimately death. There are sporadic reports of spontaneous resolution and recovery, but the natural history of this condition and the associated high rate of death is well documented.[16] Medical therapy alone has little impact on the progression of the Budd–Chiari syndrome. In a study reported by McCarthy et al.[17] 12 of 14 patients with the Budd–Chiari syndrome who were managed nonsurgically died within 6 months of diagnosis.

Table 6 provides an algorithm for the diagnosis and treatment of the Budd–Chiari syndrome. The development of ascites with or without abdominal pain should alert one to the possible diagnosis of this disorder. The presence of polycythemia, estrogen usage, previous hepatitis, or cirrhosis should heighten one's suspicion. However 19% (5 of 26) of the patients with this syndrome had no identifiable predisposing factors in our series. Altered liver function is not pronounced in the early stages of the Budd–Chiari syndrome and evaluation of serum chemistries adds little to the establishment of the diagnosis. Mild elevations of the prothrombin time and serum bilirubin may be observed, but in many patients these tests, along with the serum transaminases, are normal. Serum albumin levels may be low, reflecting both protein loss into the ascites and decreased synthesis. Hepatic venography and inferior venacavography are effective in definitively establishing the diagnosis of the Budd–Chiari syndrome. The hepatic veins may be completely occluded but frequently are visualized with the 'spider web' pattern resulting from partial recanalization.

A percutaneous liver biopsy is very useful in deciding whether one should recommend mesenteric-systemic venous decompression for these individuals. The presence of massive ascites and coagulopathy has been thought to be a contraindication to liver biopsy. In this series no morbidity or mortality resulted from routine percutaneous liver biopsies and we would suggest that the results of this procedure can significantly alter the form of surgical therapy recommended. Orthotopic liver transplantation is now an accepted surgical procedure for the treatment of end-stage liver disease and several transplant centers have advocated transplantation as the definitive therapy for the Budd–Chiari syndrome.[18] A multicenter analysis of patients with the Budd–Chiari syndrome treated by liver

TABLE 6. *Diagnosis and Treatment of Suspected Budd-Chiari Syndrome*

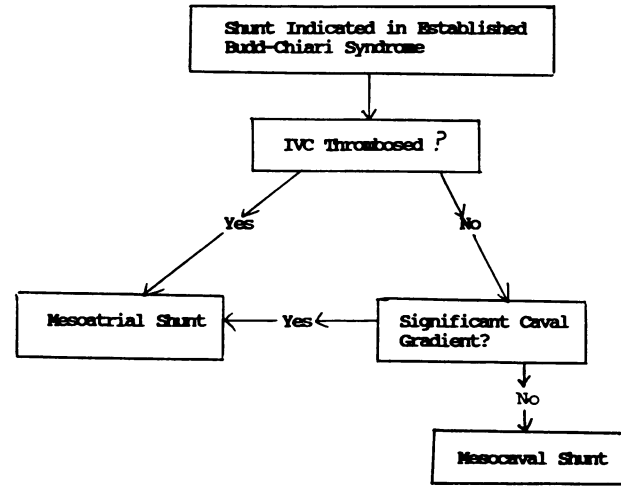


transplantation indicate a 3-year survival rate of 54%.[10] We would suggest using the liver biopsy results to determine whether a patient will benefit most from a shunt or a liver transplant. Histologically hepatic vein occlusion produces a pattern of intense centrilobular congestion. With continued obstruction to venous drainage, the hepatocytes atrophy and cell regeneration is impaired.[19] We have shown previously that following successful surgical decompression the resolution of this pathologic picture can be dramatic.[19] Liver biopsies taken several months after successful mesenteric-systemic venous shunting appear essentially normal. Sometimes, however, the liver histopathology has a significant component of cirrhosis that may be due to either protracted passive congestion of the liver secondary to hepatic vein occlusion or a preexisting illness. Laennec's cirrhosis or postnecrotic cirrhosis was identified in 15% (4) of the patients in this series. We think these patients with the Budd–Chiari syndrome with cirrhosis are best managed by orthotopic liver transplantation.

If it is decided that a portosystemic shunt is the appropriate therapy, the type of shunt must be chosen. Understanding the pathophysiology of the Budd–Chiari syndrome has allowed us to refine the surgical treatment of this illness and suggest guidelines for shunt selection. Typically the liver in patients with the Budd–Chiari syndrome appears swollen and edematous with blunted edges. The caudate lobe, which drains directly into the inferior vena cava *via* several short veins, often hypertrophies in response to the dysfunctional state of the remaining part of the organ. Frequently the caudate lobe hypertrophy produces compression of the inferior vena cava. Others have suggested that hypertrophy of the right hepatic lobe or possibly a process of pericaval fibrosis also may lead to retrohepatic caval obstruction.[5] Regardless of etiology, thrombosis or severe occlusion of the retrohepatic vena cava has technical implications for selecting the most appropriate type of portosystemic decompression. Table 7 outlines the decision tree used at the Johns Hopkins Hospital. For those patients with an established diagnosis of the Budd–Chiari syndrome who are thought to be appropriate candidates for a shunt procedure, inferior venacavography is most useful in selecting the surgical procedure. Patients with inferior vena caval thrombosis or with significant caval occlusion (compression producing a gradient more than 20 mmHg or a reduction in intraluminal diameter more than 75%) should undergo a mesoatrial shunt. Those patients without significant vena caval obstruction can be managed effectively with a mesocaval shunt.

Selection of the mesoatrial shunt for the Budd–Chiari syndrome patients with inferior vena caval compression is not a uniformly supported concept. Vons et al.[20] performed seven mesocaval shunts in patients with the Budd–Chiari syndrome who had a mean caval pressure of 15 mm of mercury. Because there were no graft thromboses in their series, they concluded that compression of the inferior vena cava should not persuade the surgeon to abandon a mesocaval or portacaval shunt in favor of a mesoatrial procedure. Based on the success of Vons and others using the mesocaval or portocaval shunts despite high caval pressures, two such patients underwent mesocaval grafting at Johns Hopkins. In one case the shunt thrombosed at 2 weeks and required conversion to a mesoatrial shunt. The second patient developed shunt stenosis at the vena caval anastomosis possibly due to continued retrohepatic caval compression and a low flow state and required balloon dilatation at 6 months. Information regarding the pressure gradient across the region of caval narrowing, the crucial factor in selecting a shunt, was not included in the study by Vons et al.[20] With the caval pressure stated in the article, the gradient could not have been as high as the 20 mm of mercury level, above which we would now favor a mesoatrial shunt.

TABLE 7. *Portosystemic Shunt Selection for Budd-Chiari Syndrome*



The perception that mesoatrial shunts are highly susceptible to thrombosis has led to a reluctance among some individuals to use this procedure. Others who have used the mesoatrial shunt have felt obliged to convert the original mesoatrial shunt to a mesocaval shunt once the hepatic congestion, and presumably caval compression, has resolved. Recently Wang et al.[21] reported 100 patients with the Budd–Chiari syndrome, 81 of whom were treated surgically in Beijing, China. Although these patients will need more extensive follow-up (range in study, 2 to 66 months) to accurately assess their outcome, 84% were alive at the time the manuscript was written. This included 22 of the 25 patients treated by mesoatrial shunt. Successful use of the mesoatrial shunt for five patients with the Budd–Chiari syndrome and concomitant vena caval occlusion or obstruction has been reported recently by Williams et

al.[22] All patients recovered and were discharged with patent shunts. Of the 15 patients in our series who underwent a mesoatrial shunt, only one developed graft thrombosis acutely. Three of the ten patients with mesoatrial shunts who were discharged alive and well from the hospital developed graft thrombosis at 2 months, 6 months, and 4 years. The last two of these patients underwent redo mesoatrial grafts using the silicone sleeve reinforced prosthesis and both patients are alive and ascites free 6 and 3 years, respectively, after their graft revision. Thus only 1 of the 10 patients developed a graft thrombosis resulting in death. Ascites is such a constant clinical component of the Budd–Chiari syndrome (more than 90%) it is unlikely that postoperative patients who remain healthy and ascites free will suffer an unrecognized graft thrombosis. We would suggest that elective conversion of a mesoatrial to a mesocaval shunt offers no clear benefit to the patient. If thrombosis does occur in the mesoatrial shunt, revision is usually successful.

In the Johns Hopkins series side-to-side portacaval shunt has not been used as a therapeutic option in the treatment of the Budd–Chiari syndrome for two reasons. The hypertrophied caudate lobe present in these patients often renders this procedure technically difficult. Furthermore, should liver transplantation be required in the future (due to shunt failure, cirrhosis, tumor, and so on), the presence of a side-to-side portacaval shunt increases the transplant mortality rate dramatically.[23] Mesocaval grafts are performed away from the porta hepatis, which then does not become a reoperative field at the time of liver transplantation. Unlike portacaval shunts, mesocaval shunts require only simple ligation at the conclusion of the transplant operation.

Although a cumulative 5-year patient survival rate of 59% with the Budd–Chiari syndrome is certainly an improvement over their predicted survival rate with medical therapy alone, the 80% long-term survival rate noted in the recently treated patients in our series is even more encouraging. Certainly a number of deaths among those patients treated early in our experience can be attributed to the moribund condition of patients selected for a procedure, which, at that time, was considered experimental. Furthermore refinements of the operative technique and graft material have decreased the risk of shunt failure. The appropriate selection of mesocaval shunt, mesoatrial shunt, or orthotopic liver transplantation should result in the safe and successful surgical management of most patients with the Budd–Chiari syndrome.

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