Results of Portal Systemic Shunts in Budd-Chiari Syndrome

CORINNE VONS, M.D.* EDWIGE BOURSTYN, M.D.* PATRICK BONNET, M.D.*
CLAUDE SMADJA, M.D.* ANNE-MARIE SZEKELY, M.D.† DOMINIQUE FRANCO, M.D.‡

Nine patients with Budd-Chiari syndrome (BCS) were treated by a portal systemic shunt. One had thrombosis of the superior mesenteric vein (SMV) and another had complete obstruction of the retrohepatic inferior vena cava (IVC). All other patients had a marked stenosis of the retrohepatic IVC with caval pressure ranging from 12 to 24 mmHg (mean: 17 mmHg). Seven patients had an interposition mesocaval shunt using an autologous jugular vein. The patient with a thrombosed SMV had a portoatrial shunt. The patient with an obstructed IVC had a cavoatrial shunt after an erroneous portacaval shunt had failed to relieve ascites. There were no operative deaths and no major postoperative complications. One patient died 19 months after operation of acute leukemia complicating polycythemia rubra vera. All other patients were alive and well 8 months to 6 years after operation. None of them had encephalopathy. These results suggest several comments: (1) Portal systemic shunts are a good treatment for BCS and have a low operative risk. (2) The mesocaval shunt is an efficient procedure, even when there is stenosis of the IVC with high caval pressure; shunts to the right atrium should be performed only in the case of complete obstruction or inaccessibility of the IVC. (3) The long-term prognosis is excellent, except in patients with potential malignancies. Therefore, portal systemic shunts should be indicated early in patients with symptomatic BCS.

From the Service de Chirurgie Hépato-Biliaire et Digestive, Service d'Anatomopathologie,† Faculté de Médecine Paris-Sud, Hôpital Paul Brousse, Villejuif, France*

IT IS WELL ACCEPTED that portal systemic shunting is an efficient treatment of Budd-Chiari syndrome (BCS) by converting the portal vein into an outflow tract, thus relieving sinusoidal congestion and preventing injury to the hepatocytes.[1-3] A variety of shunting procedures have been reported.[1,3,4] The use of the mesenteric vein seems the most appropriate since a bulky caudate lobe often prevents access to the portal vein.[5,6] Although many successful cases of shunts have now been reported, the indications for shunting in BCS are still controversial for the following reasons: (1) While the prognosis of nonoperated patients with BCS is poor, the operative mortality of shunt surgery in the reported series is high, running around 37%. (2) The most appropriate type of shunt procedure is not yet well defined. The high rate of stenosis of the inferior vena cava (IVC) behind the caudate lobe has prompted some surgeons to recommend shunts to the right atrium.[7]

Since 1978, nine patients with BCS have had a portal systemic shunt. The purpose of this paper was to analyze our results and to try to define the rationale of shunt surgery in patients with BCS.

Patients and Methods

From 1978 to 1984, nine patients with BCS have had a portal systemic shunt. During the same period of time, four patients with BCS were not operated. One had diffuse portal vein thrombosis and died of intestinal infarction before laparotomy could be undertaken. The second died of pulmonary embolism following a peritoneovenous shunt. The third had Marchiafava-Michelli disease, and decompressive surgery was contraindicated because of marked prolongation of bleeding time. He died 6 months later of ascitic infection and renal failure. The fourth patient had rapid normalization of serum transaminases, and ascites was cleared by diuretics.

The sex, age, etiological factors, chief complaint, duration of symptoms, and preoperative liver biochemical tests of the nine reported patients are summarized in Table 1. Intractable ascites and right upper quadrant (RUQ) pain were the chief complaints. In all patients, the diagnosis was confirmed by a liver biopsy obtained before operation in four cases and/or at operation in five cases.

‡ Professor at Faculté de Médecine Paris-Sud.
Reprint requests: D. Franco, M.D., Hôpital Paul Brousse, F-94804 Villejuif, France.
Submitted for publication: October 29, 1985.

TABLE 1. *Main Preoperative Features in Nine Patients with BCS*

Patient Number	Sex, Age	Etiology	Symptoms	Duration	Liver Biochemical Tests			
					ASAT (×N)	ALAT (×N)	Bilirubin (μmol/dl)	Prothrombin Time (%)
1	F, 32	Estroprogestatives	Acute ascites, RUQ pain, liver failure	3 months	5	7	5.7	44
2	F, 30	Estroprogestatives	Acute ascites, RUQ pain	6 days	70	60	1.5	28
3	M, 42	Stenosis of the ostium of hepatic veins	Intractable ascites	36 months	3	2	4.3	100
4	F, 54	Polycythemia rubra vera	Ascites, RUQ pain	7 months	8	8	3.9	50
5	F, 17	Estroprogestatives	Acute ascites, RUQ pain	1 month	3	5	3.6	53
6	F, 37	Postpartum	Intractable ascites	36 months	1	1	1.6	57
7	F, 44	Myeloproliferative disorder	Intractable ascites	14 months	1	1	2.7	46
8	F, 27	Estroprogestatives	Intractable ascites	6 months	1	1	4.6	60
9	M, 43	Not determined	Intractable ascites	60 months	1	1	2.4	85

Centrilobular necrosis and sinusoidal dilatation were the main relevant findings (Table 2). In seven patients, approximately half of each lobule was destroyed. In seven patients, the obstruction of the hepatic veins was confirmed by phlebography. There was thrombosis of the three major hepatic veins in six and stenosis of the ostium

TABLE 2. *Operative Findings in Nine Patients with BCS Treated by a Portal Systemic Shunt*

Patient Number	Aspect of the Liver	Liver Pathology	Mesenteric Vein Pressure (mmHg)	IVC Pressure (mmHg)	Pressure Gradient (mmHg)	Type of Shunt
1	Massive congestion of the left lobe, atrophy of the right lobe, hypertrophy of the caudate lobe	Massive hemorrhagic centrilobular necrosis, sinusoidal dilatation	32	12	20	Mesocaval
2	Massive congestion of the left lobe, atrophy of the right lobe, hypertrophy of the caudate lobe	Massive hemorrhagic centrilobular necrosis, sinusoidal dilatation	30	15	15	Mesocaval
3	Nodular, moderate hypertrophy	Fibrosis	30	12	18	Mesocaval
4	Congestion of the left lobe, atrophy of the right lobe, hypertrophy of the caudate lobe	Massive hemorrhagic centrilobular necrosis, sinusoidal dilatation, slight periportal regenerative hyperplasia	40	14	26	Mesocaval
5	Congestion of the left lobe, moderate atrophy of the right lobe, hypertrophy of the caudate lobe	Massive hemorrhagic centrilobular necrosis sinusoidal dilatation	22	17	5	Mesocaval
6	Massive congestion of the left lobe, atrophy of the right lobe, hypertrophy of the caudate lobe	Massive centrilobular necrosis, sinusoidal dilatation, obstruction of hepatic venules	35	24	11	Portoatrial
7	Congestion of the left lobe, atrophy of the right lobe, hypertrophy of the caudate lobe	Massive hemorrhagic centrilobular necrosis, sinusoidal dilatation	35	15	20	Mesocaval
8	Massive congestion of the left lobe, atrophy of the right lobe, hypertrophy of the caudate lobe	Massive centrilobular necrosis, sinusoidal dilatation	34	20	14	Mesocaval
9	Nodular, liver atrophy	Moderate centrilobular necrosis, sinusoidal dilatation, moderate fibrosis (sometimes annular)	24	24	0	Portacaval + cavoatrial

of the veins in one. In patient 2, a decision to proceed with life-saving decompressive surgery prevented the use of phlebography. In patient 9, thrombosis of the retrohepatic IVC when the diagnosis of BCS was established precluded hepatic phlebography.

Three patients had had one or several operations before referral. Patient 5 had had an exploratory laparotomy for acute ascites. Patient 6 had had a mesocaval shunt elsewhere that failed to relieve ascites because of thrombosis. Patient 9 was first diagnosed as having a cryptogenic cirrhosis. He underwent a Sugiura's operation because of variceal bleeding. This was followed by intractable ascites. Two successive peritoneo-jugular shunts failed because of thrombosis of the superior vena cava. Finally, he underwent a portacaval shunt. Ascites persisted despite its proven patency. The unusual course of this patient led to the assumption that the etiology of the ascites was misdiagnosed. The diagnosis of BCS was supported 5 years later by reanalyzing histopathology of the previous liver biopsy taken at the first operation.

The patency of the superior mesenteric vein, portal vein, and IVC was assessed by studying the venous phase of celiac and superior mesenteric arteriography and cavography in all patients. None of them had diffuse portal thrombosis. Patient 7 had thrombosis of the superior mesenteric vein secondary to the unsuccessful mesocaval shunt performed elsewhere. All patients had marked stenosis of the IVC on cavography. Patient 9 had thrombosis of the retrohepatic vena cava.

Eight patients had an elective procedure. Patient 2 underwent emergency surgery because of rapid increase in the serum level of transaminases and growing deterioration of liver function. The type of operation and the operative findings are summarized in Table 2. Seven patients had an interposition mesocaval shunt using an autologous (right jugular) venous graft. In patient 6, a portoatrial shunt using a polytetrafluorethylene (PTFE, Gore-Tex®) reinforced prosthesis 16 mm in diameter was performed because of thrombosis of a previous prosthetic interposition mesocaval shunt done elsewhere. Patient 9, who had already had an erroneous portacaval shunt for intractable ascites, ultimately underwent a cavoatrial shunt using a PTFE (Gore-Tex) reinforced prosthesis 16 mm in diameter.

At operation, the liver was massively enlarged and congested in 7 patients, predominantly in the left lobe. The liver was nodular in appearance in the other two patients. Portal systemic pressure gradient was measured at operation in all patients. In patient 6, it was calculated by the difference between the portal pressure measured at operation and the inferior vena caval pressure measured at preoperative cavography. The mean caval pressure was 17 mmHg (range: 12–24 mmHg). The mean pressure gradient was 10 mmHg (range: 5–25 mmHg).

All patients received heparin intravenously (1 mg per kg body weight, per day) during the first postoperative days. Three patients were given antiplatelet drugs on a long-term basis, and one with a prosthetic graft was given antivitamin K therapy.

Patient 3 was lost to follow up 2½ years after operation. All other living patients were seen within 1 month of writing this manuscript.

Results

Operative Complications

None of the patients died after operation. Patient 2, who was operated as an emergency, had a self-limited episode of encephalopathy. Liver biochemical tests rapidly improved after operation. Patient 6 had a left pleural effusion that persisted for 1 month after portoatrial shunting. In patient 7, a moderate ascites recurred after mesocaval shunt and slowly disappeared over 2 months. In the other eight patients, the ascites cleared by the end of the first postoperative month. In patients with preoperative elevation of serum transaminases, these enzymes rapidly returned to normal.

Long-term Follow-up

This is summarized in Table 3. Patient 4 died from acute myeloid leukemia 19 months after a mesocaval shunt. This was a complication of polycythemia rubra vera. Until death, she was free of ascites and had normal serum transaminases.

All other patients were alive and free of ascites when last seen. Seven of them had resumed work. Only patient 7 did not resume her work because of back pains caused by osteoporosis. Patient 1 had an uneventful pregnancy and delivery 2 years after shunting.[8] No patient showed any sign of late encephalopathy.

In almost every patient, minute changes of the liver biochemical tests persisted with a slight increase in serum bilirubin and alkaline phosphatase and a slight prolongation of the prothrombin time. Serum transaminases remained normal in all patients.

A liver biopsy was obtained in three patients 18 to 24 months after operation. There was no sinusoidal congestion. Slight fibrosis originating in the centrilobular area was observed in all three.

Discussion

Our results confirm the efficacy of portal systemic shunting in BCS: (1) it always cleared ascites when the shunt was functioning; (2) it significantly improved liver blood outflow, as demonstrated by the return to normal values of serum transaminases and the disappearance of necrosis and sinusoidal dilatation on late liver biopsies;

TABLE 3. *Late Results in Nine Patients with BCS after Portal Systemic Shunting*

Patient Number	Length of Follow-up (Months)	Present Status	Serum Bilirubin (μmol/dl)	ASAT (×N)	ALAT (×N)	Quick %	Alk. Phos. (×N)	Liver biopsy‡
1	82	Alive, no ascites, secretary, one normal pregnancy	2.0	1	1	60	1	Moderate fibrosis
2	79	Alive, no ascites professor	2.1	1	1	66*	12	Moderate fibrosis
3	30	Alive, no ascites, worker	ND	1	1	ND	1	ND
4	19	Dead from acute myeloid leukemia, no ascites at time of death.	2.8	1	1	63	2	Fibrosis, sometimes annular
5	37	Alive, no ascites, worker.	2.4	1	1	70	1	ND
6	23	Alive, no ascites, chief executive	1.8	1	1	70	1	ND
7	22	Alive, no ascites, back pain osteoporosis	4.0	1	1	60*	3	ND
8	11	Alive, no ascites, secretary	2.7	1	1	61*	2	ND
9	8	Alive, no ascites, clerk	1.6	1	1	34†	2	ND

* These three patients received antiplatelet drugs.
† This patient was treated when last seen by antivitamin K.
‡ Percutaneous liver biopsy was done 19 months after operation in patient 1, 24 months in patient 2, and 18 months in patient 4.
ND = not determined.

and (3) it dramatically reversed the acute downhill course of the patient with rapidly progressive liver failure. Three points deserve emphasis and appear particularly relevant to the indications for portal systemic shunt in BCS: (1) the absence of operative death; (2) the good results of mesocaval shunt, even in the face of a high caval pressure; and (3) the excellent prognosis on a long-term basis.

The operative mortality was nil in the present series. This favorably compares with that of other series averaging 37% (range: 16–50%).[1,3–5,7,9] A thorough analysis of the literature reveals that operative mortality is mostly related to: (1) multiple organ failure in patients operated late in the course of the disease at a time of overt hepatic insufficiency, renal failure, and poor hemodynamic conditions[3,10–12]; and (2) thrombosis of the shunt leading to recurrence of BCS and end-stage liver failure.[7,13–15] The latter complication usually occurred in two circumstances: preoperative thrombosis of the mesenteric and portal veins or IVC,[1,3] and the use of prosthetic material of small caliber.[4] A careful preoperative assessment of the patency of the superior mesenteric vein and of the IVC should prevent the performance of shunts on thrombosed veins and guide the choice of the type of shunt. It has been suggested that prosthetic grafts are more prone to thrombosis than autologous veins in portal hypertensive surgery.[16] BCS most often occurs in patients with an underlying disease that predisposes to vascular thrombosis.[2] This could enhance the risk of thrombosis of prosthetic material in those patients. Therefore, an autologous vein should be used whenever it is possible. The results of the present series suggest that when an appropriate shunt is done in a patient in a fair condition, the operative risk is quite low.

Interposition mesocaval shunt is the operation of choice in BCS since the usual enlargement of the caudate lobe precludes the construction of a portacaval shunt.[5,6] However, stenosis of the IVC and a high IVC pressure are frequently associated with the large caudate lobe, precluding the use of IVC for shunting. Shunts to the right atrium have been advocated in these circumstances.[7,17–19] Those shunts are more difficult to construct than mesocaval shunts, and they expose the patients to operative complications and late thrombosis.[2,7,9] It is worth noting that, in our series, mesocaval shunts were successful despite marked stenosis of the IVC on cavography and elevated IVC pressure in most of the patients. In patient 5, the mesocaval shunt was functioning, although caval pressure was 17 mmHg and the mesocaval pressure gradient was 5 mmHg. Therefore, shunts to the right atrium should be reserved for patients with (1) complete obstruction of the IVC as in our case 9, (2) no pressure gradient between the mesenteric vein and the IVC, or (3) an inaccessible IVC as in our case 6. Interposition mesocaval

shunt with a venous graft should always be favored in other cases.

Long-term results were quite satisfactory in the present series. The only death occurred in a patient with polycythemia rubra vera and was due to malignant transformation. All other patients had a normal life. These results would suggest that apart from patients with malignant etiologies of their BCS the postoperative life expectancy is good. Two reservations, however, should be noted. (1) Liver biochemical tests in these patients did not completely return to normal values, and the evolution may progress slowly towards liver failure. (2) It has recently been mentioned that primary myeloproliferative disorder without overt changes in peripheral blood was frequent in patients with hepatic vein thrombosis.[20] If this is confirmed, late prognosis might depend on the evolution of such minor myeloproliferative disorders.

The natural history of BCS is still uncertain, and the results of the published series are difficult to evaluate. However, when ascites or continuing liver necrosis is present, life expectancy is dismal.[21] In the series of Tavill et al., six of 19 patients with BCS died within the first year and 17 were dead after 3.5 years.[22] The unfavorable evolution of our three nonoperated symptomatic patients would emphasize those figures. The high reported operative mortality after portal systemic shunting may have discouraged wide application of shunt surgery in BCS. Liver transplantation has even been advocated in several cases.[23] Our results suggest that portal systemic shunts are a safe operation in patients with BCS. Millikan et al. suggested that surgery should be indicated in patients with severe necrosis of the central area of the lobules.[7] We strongly believe that a portal systemic shunt should be urgently performed in a patient with a symptomatic BCS and patent portal vessels, irrespective of any other consideration. Mesocaval shunting with an autologous venous graft is the operation of choice. Shunts to the right atrium should be performed only in the few cases in which the former procedure is not feasible.

Acknowledgments

The authors are very grateful to D. Valla and O. Traynor for carefully revising this manuscript.

References

1. Langer B, Stone RM, Colapinto RF, et al. Clinical spectrum of the Budd-Chiari syndrome and its surgical management. Am J Surg 1975; 129:137–145.
2. Mitchell MC, Boitnott JK, Kaufman S, et al. Budd-Chiari syndrome: etiology, diagnosis and management. Medicine 1982; 61:199–218.
3. Orloff MJ, Johansen KH. Treatment of Budd-Chiari syndrome by side-to-side portacaval shunt: experimental and clinical results. Ann Surg 1978; 188:494–510.
4. Huguet C, Liegeois A, Levy VG, Caroli J. Interposition mesocaval shunt for chronic primary occlusion of the hepatic veins. Surg Gynecol Obstet 1979; 148:691–698.
5. McCarthy PM, Van Heerden JA, Adson MA, et al. The Budd-Chiari syndrome: medical and surgical management of 30 patients. Arch Surg 1985; 120:657–662.
6. Cameron JL, Maddrey WC. Mesoatrial shunt: a new treatment for the Budd-Chiari syndrome. Ann Surg 1978; 187:402–406.
7. Millikan WJ, Henderson JM, Sewel CW, et al. Approach to the spectrum of Budd-Chiari syndrome: which patients require portal decompression? Am J Surg 1985; 149:167–176.
8. Vons C, Smadja C, Franco D, et al. Successful pregnancy in Budd-Chiari syndrome. Lancet 1984; 2:975.
9. Cameron JL, Herlong HF, Sanfey H, et al. The Budd-Chiari syndrome: treatment by mesenteric systemic venous shunts. Ann Surg 1983; 198:335–344.
10. Schramek A, Better OS, Brook JG, et al. New observations in the clinical spectrum of the Budd-Chiari syndrome. Ann Surg 1974; 180:368–372.
11. Dykes PW, Ellis HA, Gourevitch A. The Budd-Chiari syndrome in a patient with hepatic gummata. Br J Surg 1963; 227:934–937.
12. Ludwich JR, Markel SF, Child CG III. Chiari's disease: a clinicopathological study. Arch Surg 1965; 91:697–704.
13. Hoyumpa AM, Schiff L, Helfman EL. Budd-Chiari syndrome in women taking oral contraceptives. Am J Med 1971; 50:137–140.
14. Hautefeuille P, Lecharny B, Nusinovici V, et al. Anastomose porto-cave latéro-latérale avec greffon veineux interposé dans 2 cas de syndrome de Budd-Chiari chronique. Nouv Presse Med 1974; 3: 1104.
15. Meindock H, Langer B. Liver scan in Budd-Chiari syndrome. J Nucl Med 1975; 17:365–368.
16. Haimovici H, Hoffert PW, Zinicola, Steinman C. An experimental and clinical evaluation of grafts in the venous system. Surg Gynecol Obstet 1970; 131:1173–1186.
17. Warren WD, Potts JR, Fulenwider JT, et al. Two stage surgical management of Budd-Chiari syndrome associated with obstruction of the inferior vena cava. Surg Gynecol Obstet 1984; 159: 101–107.
18. Wruble LD, Scott JC, Wolf RY. Budd-Chiari syndrome: dramatic relief with mesoatrial shunt. Am J Gastroenterol 1982; 77:253–255.
19. Khanduri P, Murlidharan, Nathaniel P, et al. Mesoatrial shunt in the surgical treatment of Budd-Chiari syndrome. Aust NZ J Surg 1983; 53:309–315.
20. Valla D, Casadevall N, Lacombe C, et al. Primary myeloproliferative disorder and hepatic vein thrombosis: a prospective study of erythroid colony formation in vitro in 20 patients with Budd-Chiari syndrome. Ann Intern Med 1985; 103:329–334.
21. Parker BGF. Occlusion of the hepatic veins in man. Medicine 1959; 38:369–402.
22. Tavill AS, Wood EJ, Kreel L, et al. The Budd-Chiari syndrome: correlation between hepatic scintigraphy and the clinical, radiological, and pathological findings in nineteen cases of hepatic venous outflow obstruction. Gastroenterology 1975; 68:509–518.
23. Seltman HJ, Dekker A, Van Thiel DH, et al. Budd-Chiari syndrome recurring in a transplanted liver. Gastroenterology 1983; 84:640–643.