The Budd-Chiari Syndrome

Treatment by Mesenteric-Systemic Venous Shunts

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Twelve patients with the Budd-Chiari syndrome have been managed surgically. Ten of the patients were female, two were male, with a mean age of 40 years. Three of the patients had polycythemia vera, two had pre-existing cirrhosis, one had ingested estrogens, one had an occult tumor, and in four there were no associated factors. Ten patients presented with ascites and two with bleeding esophageal varices. The diagnosis was confirmed in all 12 patients by liver biopsy and hepatic vein catheterization. Inferior vena cavography revealed the abdominal vena cava to be thrombosed in six patients. The superior mesenteric vein was used to decompress the congested liver in all 12 patients. In five patients, a mesocaval shunt (MCS) was performed and in seven patients, a mesoatrial shunt (MAS) was carried out. There were four hospital deaths (two MCS, two MAS). One late death (MAS) occurred from liver failure following shunt thrombosis. Two additional patients (one MCS, one MAS) redeveloped ascites immediately following surgery and angiography revealed a thrombosed shunt. Ascites has been controlled with a LeVeen shunt in these two patients, but liver biopsies showed progression to cirrhosis. The remaining five patients (three MAS, two MCS) did well, and angiography revealed patent shunts. Two of these patients, however, redeveloped ascites at 4 and 10 months following MAS and required a second MAS. Follow-up ranges from 6 to 68 months. In three of the patients (two MCS, one MAS) with patent shunts, liver biopsy shows a remarkable return toward normal liver architecture and histology.

THE BUDD-CHIARI SYNDROME is an unusual form of portal hypertension resulting from occlusion of the major hepatic veins. When the hepatic veins become occluded, the centrilobular regions of the liver become intensely congested. Cell atrophy and impaired regeneration result. Patients with this entity usually present with larger tender livers and with massive ascites secondary to a markedly congested liver. When patients present acutely, the syndrome is rapidly fatal unless treated surgically. A more chronic form of the disease results in portal hypertension with esophageal variceal hemorrhage.[1,2] Portasystemic shunting operations have been effective in managing patients with the Budd-Chiari syndrome by converting the portal vein into an outflow tract, thus decompressing the massive hepatic congestion. Side-to-side portacaval shunts,[3] splenorenal shunts,[4] and mesocaval shunts[5] have all been used successfully for this purpose. For some patients, extension of thrombus into the inferior vena cava makes traditional shunting procedures impossible.[6] Our recent experience suggests that vena caval occlusion frequently complicates hepatic vein occlusion. Our experience with 12 patients with the Budd-Chiari syndrome treated surgically is reported. In all patients, the superior mesenteric vein was used as the outflow tract to decompress the liver. If the inferior vena cava was patent, a mesocaval shunt was performed. If the inferior vena cava was occluded, a mesoatrial shunt was carried out.

Clinical Material

Between 1973 and 1983, 12 patients with the Budd-Chiari syndrome were managed surgically at The Johns Hopkins Hospital (Table 1). Seven of the 12 patients have been treated since 1981. Ages have ranged from 14 to 64 years, with a mean of 40 years. Ten of the patients were female and two were male. Nine of the patients were white, two were black, and one was Cambodian. Ten of the 12 patients presented with massive ascites. One patient presented with ascites and bleeding esophageal varices and the final patient presented with repeated

Presented at the Annual Meeting of the American Surgical Association, May 12–14, 1983, Boca Raton, Florida.

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Submitted for publication: May 16, 1983.

0003-4932/83/0900/0335 $01.40 © J. B. Lippincott Company

TABLE 1. *Patients Undergoing Mesenteric-Systemic Venous Shunts for the Budd-Chiari Syndrome*

Patient	ARS	Presentation	Duration of Symptoms	Associated Disease	Admission Liver Function Tests						Inferior Vena Cavography	Shunt Procedure Date	Ascites	Uncorrected Portal Pressure Pre and Post Shunt cmH_2O	Follow-up
					BIL mg/dl	SGOT IU/L	SGPT IU/L	ALK Phos IU/L	ALB g/dl	Serum Creat mg/dl					
1 SG	36 WF	Ascites jaundice	8 mo.	Estrogen ingestion	Deeply jaundiced						Clotted	Mesoatrial 6/73	8 L	29–14	Died 2 days postop: liver failure
2 TE	14 BF	Ascites abdominal pain	2 yr.	None	0.8	50	22	146		0.6	Patent	Mesocaval 4/76	1 L	24–16	Shunt thrombosed LeVeen shunt—cirrhosis
3 CS	57 BF	Esophageal bleeding	4 yr.	Alcoholic cirrhosis	0.6	25	14	106	4.0	0.6	Clotted	Mesoatrial 10/76	1.5 L	30–10	Survived 5 yrs: no ascites, no bleeding
4 AG	44 WF	Ascites	1 mo.	Polycythemia vera	3.0	22	14	102	3.6	0.8	Patent elev. press.	Mesoatrial 3/77	9 L	38–15	Shunt thrombosed: LeVeen shunt: liver fibrosis
5 AL	28 WF	Ascites	4 mo.	None	2.4	17	7	88	3.2	0.5	Patent	Mesocaval 9/77	1.5 L	45–28	Shunt patent: doing well 5 yrs., 8 mos.
6 DB	33 WM	Ascites	6 mo.	Paroxysmal nocturnal hemoglobinuria	2.0	78	24	120	3.6	1.0	Patent	Mesocaval 1/81	3 L	41–28	Shunt patent: doing well 2 yrs., 4 mos.
7 JO	40 WF	Ascites abd. pain	1 yr.	Polycythemia vera	2.7	41	14	272	3.6	1.5	Clotted	Mesoatrial 2/82	26 L	35–17	Shunt stenosis at 10 mos, shunt redone: doing well 15 mos.
8 EC	64 WF	Ascites	6 mo.	Polycythemia vera	1.5	17	9	165	3.3	2.6	Patent	Mesocaval 6/82	4 L	30–11	Died 17 days postop of hepatic and renal failure
9 HS	60 WF	Ascites jaundice	4 mo.	None	7.1	33	15	134	3.2	2.6	Patent	Mesocaval 7/82	5 L	40–16	Died 14 days postop of hepatic and renal failure
10 SN	27 AM	Ascites esophageal bleeding	1 yr.	Post necrotic cirrhosis	1.2	39	16	164	3.4	1.0	Clotted	Mesoatrial 11/82	1 L	50–25	Shunt thrombosed at 4 mos.: shunt redone: doing well 6 mos.
11 TZ	28 WF	Ascites abd. pain	2 mo.	None	0.7	44	40	326	3.0	1.2	Clotted	Mesoatrial 11/82	5 L	50–25	Portal system thrombosed at 2 mos.: LeVeen shunt: died 2 mos. later of liver failure
12 VL	42 WF	Ascites abd. pain	5 mo.	Occult lung cancer cardio-myopathy	1.1	118	103	312		0.7	Clotted	Mesoatrial 2/83	10 L	35–25	Died 16 days postop of cardiac and renal failure

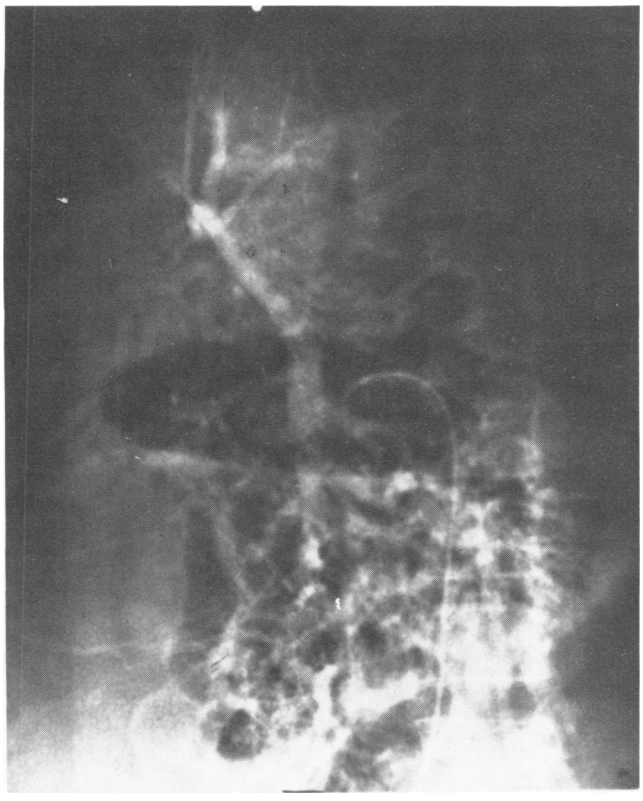

FIG. 1. Mesenteric angiogram of a 60-year-old woman (HS) with the Budd-Chiari syndrome. The diameter of the superior mesenteric vein is greater than that of the portal vein, presumably secondary to reversed flow.

esophageal bleeding without obvious ascites. At the time of surgery, however, ascites were present. Duration of symptoms prior to admission to Hopkins in the 11 patients with clinical ascites varied from 1 month to 2 years and averaged 8 months. The final patient had had repeated episodes of esophageal bleeding over a 4-year period. Although abdominal discomfort was experienced by all 12 patients, only four patients complained specifically of abdominal pain on admission. Two patients were clinically jaundiced at presentation. Three patients had polycythemia vera, and this was presumed to predispose to hepatic vein thrombosis. Two patients had pre-existing cirrhosis. In one patient, the cirrhosis was secondary to alcohol ingestion and in the other, it was thought to be chronic active hepatitis B. One patient had a 4-year history of paroxysmal nocturnal hemoglobinuria. One patient had a history of estrogen ingestion 6 years prior to the onset of her ascites. At autopsy, one patient was found to have an occult squamous cell carcinoma of the lung with tumor cells contained in the clot in the inferior vena cava. This patient also had a cardiomyopathy. In the remaining four patients, there were no predisposing factors known to be associated with the Budd-Chiari syndrome. On admission, 11 of the 12 patients had ascites, usually massive. Marked hepatomegaly was present in all 12 patients. In three patients, leg, thigh, and lower trunk edema was present, suggesting inferior vena cava occlusion. In all patients, a prominent venous pattern was present on the abdominal wall. Liver function tests were remarkably normal on admission in 11 patients (Table 1). Mean serum bilirubin was 2.1 mg/100 dl (range, 0.6–7.1 mg/100 dl), mean SGOT was 44 IU/L (range, 17–118 U/L), mean SGPT was 25 IU/L (range, 7–103 IU/L), and mean serum alkaline phosphatase was 176 IU/L (range, 88–326 IU/L). One patient (SG) was deeply jaundiced at the time of surgery. The diagnosis of the Budd-Chiari syndrome was confirmed in all 12 patients by liver biopsy and catheterization studies. Liver histology will be discussed later. In all 12 patients, hepatic vein catheterization demonstrated either the typical recanalized "spider web" pattern, or else the completely occluded veins could not be engaged with the catheter. Superior mesenteric arteriography demonstrated the portal and superior mesenteric veins to be patent in all 12 patients. A characteristic pattern, presumably secondary to reversed flow, of the superior mesenteric vein having a larger diameter than the portal vein, was seen often (Fig. 1). Inferior vena cavography demonstrated the inferior vena cava to be clotted in six of the 12 patients (Fig. 2). In one additional patient, the inferior vena cava pressure was elevated to 25 mmHg, secondary to caval compression by the hypertrophied caudate lobe in the absence of clot. In the remaining five patients, despite the frequent venographic appearance of vena caval compression, the inferior vena caval pressure was normal. Eight of the 12 patients had Technitium[99] liver scintiscans and in three patients the characteristic area of central uptake in the hypertrophied caudate lobe

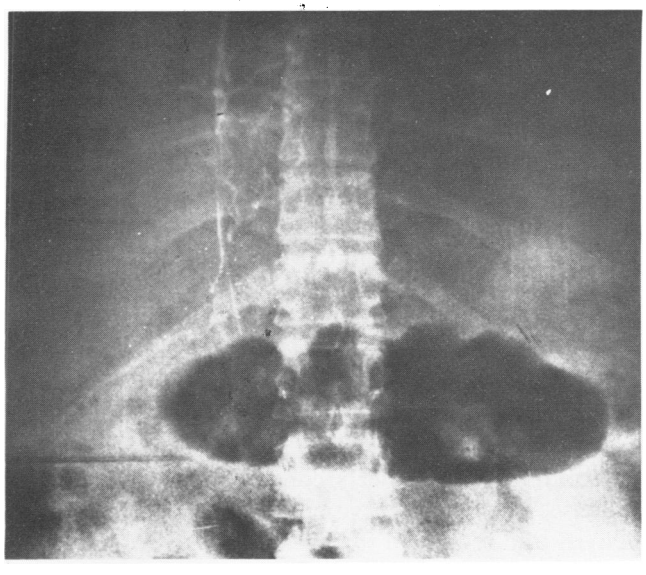

FIG. 2. Inferior vena cavagram of a 40-year-old woman (JO) with the Budd-Chiari syndrome. The study demonstrates a clotted vena cava with contrast media seen around the thrombus.

was seen. One patient (SG) at the time of surgery was in hepatic coma, and two additional patients (EC, HS) had both hepatic and renal failure.

Operative Management

Five patients underwent mesocaval interposition shunts. All five patients had inferor vena cavograms showing that patent vessel, and pressures were within the normal range. An 18-mm knitted Dacron® graft was used in four patients. In the fifth patient (TE), a rigid 16-mm woven Dacron graft was used. The graft was anastomosed end to side to the inferior vena cava and then brought anteriorly below the third portion of the duodenum. The graft was then swung up anterior to the duodenum and on top of the uncinate process of the pancreas and anastomosed obliquely to the anterior surface of the superior mesenteric vein just before it passed posterior to the neck of the pancreas. This modification has been described previously as the mesocaval "C" shunt.[7] The amount of ascites removed from the five patients at the time of surgery ranged from 1 to 5 liters. The liver was markedly congested in all five patients. The uncorrected mean portal venous pressure (Table 1) pre shunt was 36 cmH_2O (range 24–45 cmH_2O), and post-shunt pressure fell to 20 cmH_2O (range, 11–28 cmH_2O).

The remaining seven patients underwent mesoatrial shunts. Angiograms demonstrated the inferior vena cava to be thrombosed in six patients. The seventh patient (AG) had a patent inferior vena cava but with an elevated pressure (25 mmHg) secondary to compression by a hypertrophied caudate lobe. A 16-mm woven Dacron graft was used. The graft was anastomosed end to side to the superior mesenteric vein just before the vein passed posterior to the neck of the pancreas. The graft was then passed through the transverse mesocolon, anterior to the stomach, and under the xiphoid process into the superior mediastinum. The graft was then passed into the right chest and, through a separate anterior thoracotomy, was anastomosed end to side to the right atrium. The procedure has been described in detail previously.[6] The amount of ascites removed at the time of surgery in patients undergoing mesoatrial shunts ranged from 1 to 26 liters. The liver was markedly congested in all seven patients. In addition, two patients had macronodular cirrhosis. The uncorrected mean portal pressure (Table 1) pre shunt was 38 cmH_2O (range, 29–50 cmH_20), and post-shunt pressure fell to 19 cmH_2O (range, 10–25 cmH_2O).

Clinical Course

Two of the five patients undergoing mesocaval shunts died after operation. One patient (EC) died 17 days following surgery of renal and hepatic failure. Autopsy permission was not granted. The second death (HS) occurred 14 days following surgery and was also secondary to renal and hepatic failure. Post-mortem examination demonstrated the shunt to be patent. These two patients were 64 and 60 years of age, respectively, and both had elevated serum bilirubin and creatinine levels prior to surgery. Three patients tolerated surgery well and were discharged from the hospital. Two patients (DB, AL) have remained free of ascites and have angiographically demonstrated patent shunts. They are 2 years and 4 months and 5 years and 8 months post-mesocaval shunts. Postoperative liver biopsies in both patients show a remarkable return toward normal architecture and histology (Fig. 3). The final patient redeveloped ascites after operation, and angiography demonstrated a thrombosed shunt. Her ascites has been effectively controlled with a LeVeen shunt. A recent liver biopsy shows a well established cirrhosis.

Among the seven patients undergoing a mesoatrial shunt, there were two postoperative deaths. One patient (SG) was deeply jaundiced and in hepatic coma at the time of surgery. She died two days following mesoatrial shunt. Autopsy permission was not granted. The second patient (VL) died 16 days following surgery of cardiac and renal failure. She was felt to have a restrictive cardiomyopathy that was not recognized before operation. Because of a wound dehiscence, she was re-explored 6 days following surgery. The graft was patent, the liver was decompressed, and a liver biopsy showed marked improvement (Fig. 4). Post mortem examination following her death 10 days later revealed an occult lung cancer and tumor cells in the thrombus occluding her inferior vena cava. The remaining five patients tolerated surgery well and were discharged from the hospital. One patient (CS) remained free of ascites and esophageal bleeding for 5 years before succumbing to an unrelated illness. Postoperative angiography demonstrated her shunt to be patent. A second patient (JO) redeveloped ascites 10 months after surgery. Initial postoperative angiography had demonstrated a patent shunt. When her ascites recurred, repeat angiography demonstrated the shunt to be patent but compressed where the graft passed beneath the xiphoid. She was reoperated on and her graft was replaced with a 16-mm woven Dacron graft with a silastic cuff positioned so that graft compression by the xiphoid could not occur. Postoperative angiography demonstrated the graft to be patent. She is now 15 months following her first mesoatrial shunt and 5 months following her second mesoatrial shunt, and is ascites free. Another patient (SN) returned to normal activity and remained free of ascites for 4 months. The initial postopertive angiogram demonstrated his shunt to be open. Ascites then recurred and angiography demonstrated the graft to be occluded. The patient was re-

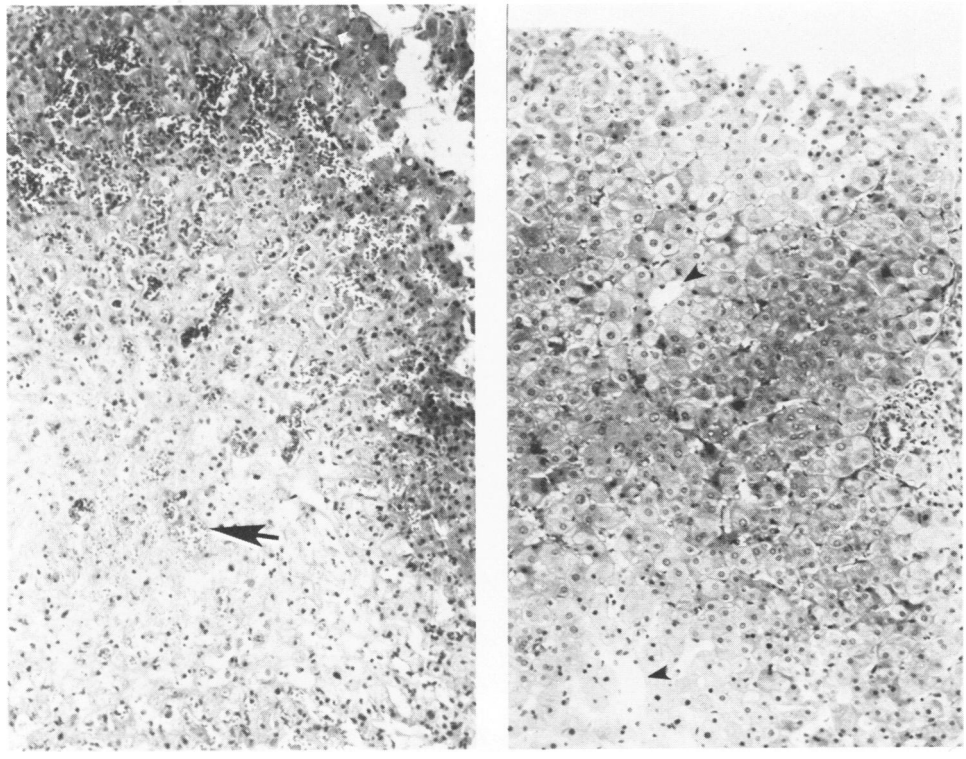

FIG. 3. Photomicrographs of liver biopsy specimens from patient AL. A, preoperative biopsy: there is extensive hepatocellular loss, involving at least 50% of the lobule, around a small terminal hepatic venule (arrow). Adjacent parenchyma is congested intensely (hematoxylin and eosin stain, ×125). B, biopsy taken eight months following successful mesocaval shunt. Congestion and hepatocellular loss are no longer evident. Arrows point to small terminal hepatic venules (hematoxylin and eosin stain, ×125).

operated on, the old graft removed, and a new 16-mm Goretex graft with external support rings inserted. The patient did well and angiography demonstrated the shunt to be patent 1 month following the operation. He is now back to normal activity 6 months following the initial operation and 2 months following his second operation.

In one patient (TZ), ascites disappeared following surgery and angiography demonstrated a patent shunt. However, 2 months later, abdominal pain and ascites suddenly developed. Angiography and laparotomy demonstrated that the entire portal system was thrombosed. Ascites in this patient was managed with a LeVeen shunt, but she died two months later of liver failure. In one patient (AG), ascites recurred during the immediate postoperative period and angiography demonstrated a thrombosed shunt. Her superior mesenteric vein was very small at the time of surgery, and the decision was made not to reoperate on her. Her ascites has been controlled effectively with a LeVeen shunt. Liver biopsy demonstrates continued congestion and fibrosis.

Pathology

Twenty-seven needle or wedge liver biopsies were available from these 12 patients, seven of which were taken following surgical decompression. The hepatic histopathology covered the full spectrum reported in the Budd-Chiari syndrome which, aside from the hepatic vein thrombosis, is basically that of severe passive congestion. Sinusoids were consistently dilated in the centrolobular (peripheral acinar) areas but contained highly variable numbers of blood cells. In association with this, there was hepatocellular atrophy progressing, in almost all of our cases, to actual loss of hepatocytes in these regions (Figs. 3, 4). Extravasation of red blood cells into the space of Disse, which in its most dramatic form leads to a picture of empty dilated sinusoids with closely packed red blood cells filling the hepatic cords and replacing lost hepatocytes, was evident in most cases but prominant in only a few. Despite the sometimes considerable loss of hepatocytes, necrotic cells were seldom numerous and inflammation was not a significant feature; large areas of frank ischemic necrosis were seen only in conjunction with superimposed portal vein thrombosis or systemic hypotension. While in protracted cases, fibrosis and eventually true cardiac type cirrhosis may develop, the picture of severe congestion and hepatocellular loss without significant fibrosis or evidence of hepatocellular rgeneration was seen even after many months of clinical illness. Thus, although there was an apparent progressive loss of centrolobular hepatocytes in several cases in which multiple preoperative biopsies were done, there were no qualitative features that were clearly related to duration of the disease.

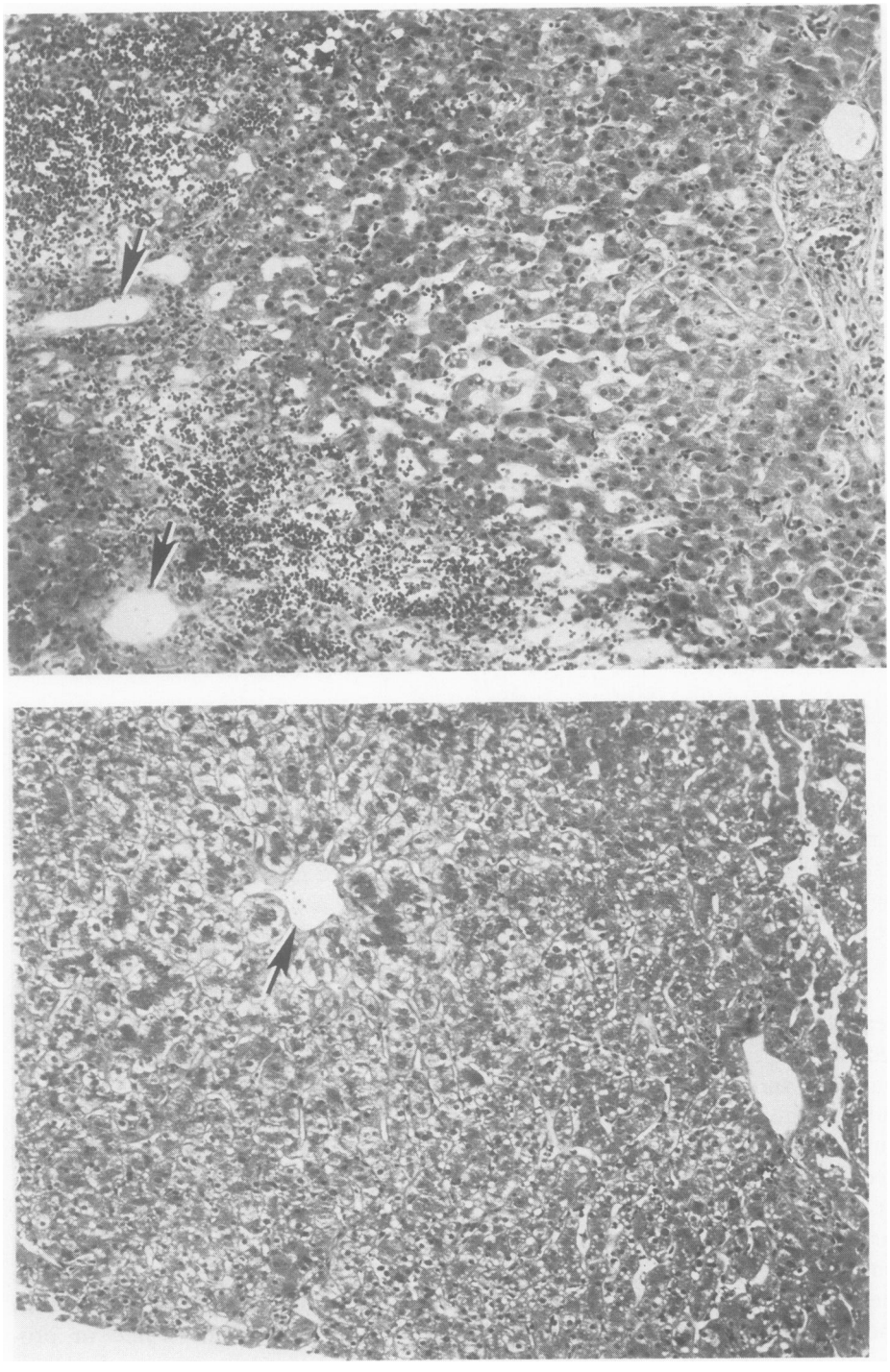

FIG. 4. Photomicrographs of liver biopsy specimens from patient VL. A, intraoperative biopsy showing intense congestion with apparent loss of hepatocytes adjacent to small hepatic veins (arrows). Sinusoidal dilatation is evident in the mid-zonal area. Note normal portal triad in upper right corner (Hematoxylin and eosin stain, ×125). B, biopsy taken six days following mesoatrial shunt. There is a moderate small droplet fat accumulation, but congestion and cell loss are no longer evident and the centrolobular area has been repopulated by slightly swollen hepatocytes. Arrow points to small hepatic vein; a small portal triad is at the right edge of this photomicrograph (Hematoxylin and eosin stain ×125).

Upon successful surgical decompression, the histologic picture changed dramatically and rapidly (Figs. 3, 4). Sinusoidal dilatation and congestion markedly decreased in most areas, although a few foci of residual congestion were occasionally apparent. Repopulation of previous areas of cell loss was prominant and, in one case that was re-biopsied only 6 days following a mesoatrial shunt, was already essentially complete (Fig. 4). Most patients did show some small areas of centrolobular collapse and fibrosis, but even when this was sufficient to suggest progression towards cirrhosis, the diminished congestion and hepatocellular regeneration were associated with clinical improvement, and the functional significance of this is doubtful.

Discussion

Occlusion of all three major hepatic veins results in massive liver congestion with the formation of large quantities of ascitic fluid. This entity, recognized for over 100 years,[8] is referred to as the Budd-Chiari syndrome. The initial clinical presentation can be benign, but eventually portal hypertension develops with esophageal varices and liver failure.[1,2] In this country, the disease invariably results from occlusion of the major hepatic veins. Although the occlusion can be secondary to tumor, most commonly hepatoma, adrenal cell carcinoma, and hypernephoma, in most instances the veins are thrombosed.[1,2] The pathogenesis of the hepatic vein thrombosis is not known, but associated diseases and factors have been identified. Polycythemia vera, paroxysmal nocturnal hemoglobinuria, oral contraceptive use, and pregnancy are the most commonly associated conditions. All of these conditions are associated with an increased incidence of venous thrombosis. It is interesting that two of our 12 patients had pre-existing cirrhosis, an association not noted in the past with the Budd-Chiari syndrome with any frequency. A recent review has demonstrated that 30% of cases are idiopathic.[1] This contrasts to an extensive review published in 1959 in which 70% of cases had no associated disease.[2] In the Far East, the Budd-Chiari syndrome has been described secondary to membranous webs partially occluding the inferior vena cava above the hepatic veins.[9] Such webs are extremely rare in this country. Membranotomy may be effective therapy in these patients.

The natural history of the Budd-Chiari syndrome is not known accurately. In some patients, the ascites may resolve spontaneously, and the patient may survive indefinitely. However, most studies and reviews suggest that the majority of patients with this disease will succumb to liver failure or bleeding if not treated surgically.[1-3] The medical management of the Budd-Chiari syndrome consists of diuretic administration, anticoagulation, and thrombolytic therapy and has largely been ineffective.[1] Diuretics alone rarely control ascites. Anticoagulants may be helpful in preventing extension of the thrombotic process to involve the inferior vena cava or portal vein but do not dissolve the hepatic vein thrombi. Several patients have been treated with streptokinase with disappearance of ascites.[1,10,11] However, treatment with this agent ideally should be initiated within a week of the thrombosis, and in most patients, the diagnosis is not made for several weeks—even months.

Surgical approaches to the Budd-Chiari syndrome have been varied. Some have advocated peritoneovenous shunting to control the ascites,[12] assuming the hepatic veins will recanalize and hepatic function return to normal. Although this approach will undoubtedly control ascites, as demonstrated by three patients in our series (AG, TE, TZ), it is unlikely to have any beneficial long-term effect on the liver. One of the three patients (TZ) in our series, managed with a LeVeen shunt, died of liver failure; another patient (TE) subsequently developed a well established cirrhosis; the third patient (AG) has continued congestion and fibrosis. All three of these patients had previously thrombosed their mesenteric-systemic venous shunt. Another approach has been that of Akita and Sakoda,[13] decompressing the liver via portopulmonary shunts established by splenopneumopexy. However, this operation, which establishes an area of contact between the abraded spleen and the lung through a defect in the diaphragm, can only be performed after ascites has been controlled. In this country, most patients with the Budd-Chiari syndrome have refractory ascites, and thus are not candidates for this operation. Liver transplantation has also been performed in at least three patients with the Budd-Chiari syndrome,[14,15] with two of three patients being alive after 1 year.

Perhaps the oldest, and clearly the most effective means of managing the Budd-Chiari syndrome surgically is to decompress the congested liver by converting the portal vein into an outflow tract. This can be accomplished by any of the functional side-to-side portasystemic shunts. The first patient successfully treated in this manner was reported by Blakemore in 1948.[16] The side-to-side portacaval shunt has been used most frequently, and a recent review documented 27 patients treated by this operation.[1] Nine patients survived the procedure and were clearly benefited. The same review reported ten patients who underwent conventional splenorenal shunt for the Budd-Chiari syndrome with three successes. Seven of nine patients in the review[1] undergoing mesocaval shunts did well. The efficacy of converting the portal vein into an outflow tract in managing the Budd-Chiari syndrome was demonstrated conclusively by Orloff and Johansen[3] in an experimental and clinical study. In a dog model of the Budd-Chiari syndrome, survival was clearly increased and liver histology improved by a side-to-side portacaval shunt. Improvement in liver histology, as well as long-term survival, was also documented in five of six patients with this disease undergoing side-to-side portacaval shunt.

In managing the Budd-Chiari syndrome, our preference has been to use the superior mesenteric vein, rather than the portal vein, to decompress the liver. Surgical access to the portal vein can be difficult. The liver in this disease is massively congested and markedly enlarged. In addition, the caudate lobe usually is hypertrophied. The caudate lobe venous drainage is directly into the inferior vena cava via several small veins that

often are not involved by the thrombotic process and thus the caudate lobe hypertrophies. This can displace both the portal vein and inferior vena cava, making apposition of the two venous structures difficult, or even impossible (Fig. 5). A mesocaval "C" shunt is performed below the transverse mesocolon, well away from the congested liver and hypertrophied caudate lobe, and can be performed with ease in the Budd-Chiari syndrome (Fig. 6).

Unfortunately, not all patients with the Budd-Chiari syndrome are candidates for a mesocaval shunt. In some patients, the thrombotic process also involves and occludes the inferior vena cava, eliminating the standard portasystemic shunts from use (Fig. 2). The frequency of this complication is not known, but of the six patients operated on at Hopkins with the Budd-Chiari syndrome since 1982, four have had a thrombosed inferior vena cava. In addition, another patient (MH:JHH# 203 02 65) was admitted to Hopkins in September of 1982 with the Budd-Chiari syndrome and a clotted inferior vena cava. Two days after admission, while being prepared for surgery, she died suddenly. An autopsy demonstrated a massive pulmonary embolus. Thus, five out of seven patients seen over the past 2 years had a thrombosed inferior vena cava. The mesoatrial shunt was first reported in 1978[5] as a treatment for the Budd-Chiari syndrome when the inferior vena cava is occluded (Fig. 7). Other reports have subsequently appeared,[17,18] and a recent literature review reported four successes out of seven patients.[1]

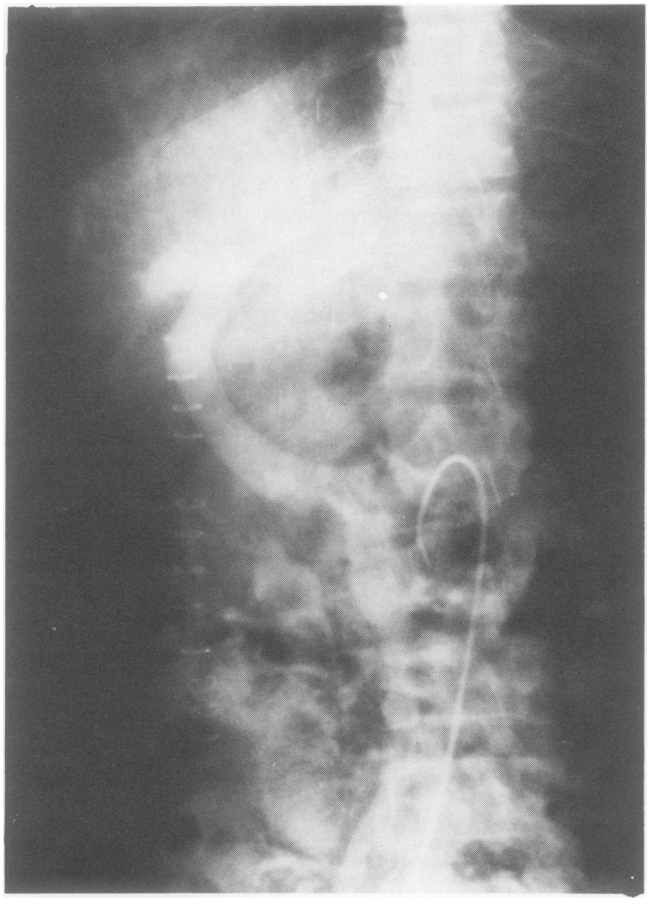

FIG. 5. Mesenteric angiogram of a 33-year-old man (DB) with the Budd-Chiari syndrome. The portal vein can be seen to be markedly displaced laterally by a hypertrophied caudate lobe.

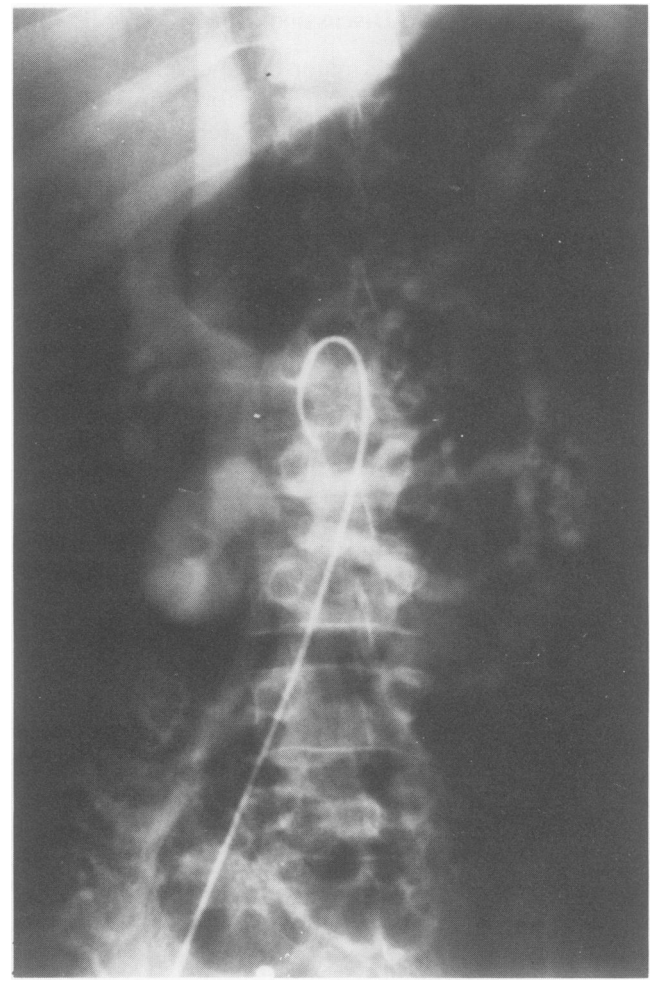

FIG. 6. Mesenteric angiogram of a patient with the Budd-Chiari syndrome (DB, Fig. 5) following a mesocaval "C" shunt. Contrast media can be seen outlining the "C" Dacron® shunt and opacifying the inferior vena cava.

Not all patients in our series were managed successfully. Two of the five patients undergoing a mesocaval shunt died after operation. Both were 60 years of age or older, old for the Budd-Chiari syndrome, and both had elevated serum bilirubin and creatinine levels prior to surgery. Neither patient tolerated the operative procedure, and both died of renal and hepatic failure. In one of the three survivors, the mesocaval shunt thrombosed. This patient (TE) was 14 years old and had a

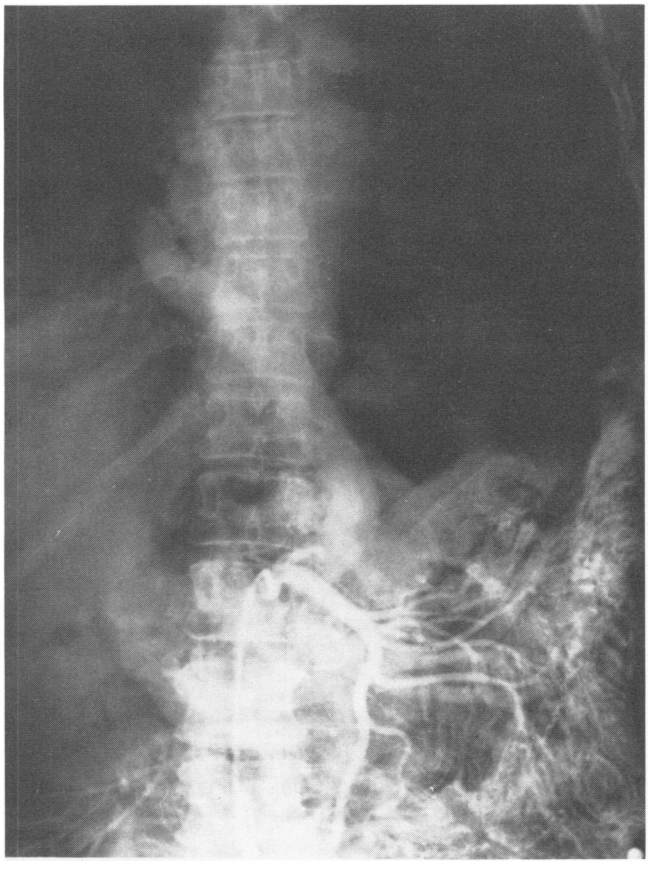

FIG. 7. Mesenteric angiogram of a 57-year-old woman following a mesoatrial shunt. Contrast media can be seen outlining the shunt and opacifying the right atrium.

small delicate superior mesenteric vein. A rigid woven Dacron graft was used for the shunt, and this resulted in distortion of the vein. Thus, this failure was secondary to technical features. The remaining two patients are long-term survivors, and liver biopsies show a marked improvement in histology.

Two patients died after operation following mesoatrial shunts. One patient (SG) had severe hepatic failure at the time of surgery and the other (VL) had a restrictive cardiomyopathy not recognized before surgery. Neither patient tolerated surgery and both died in the postoperative period. It is interesting that one of these patients (VL) dehisced her abdominal wound at six days following surgery and had to be reoperated on. At surgery, her shunt was open, her liver had decreased in size, and a liver biopsy showed almost complete hepatocyte repopulation even at this early stage of decompression (Fig. 4). Of the remaining five patients undergoing mesoatrial shunts, one thrombosed immediately following surgery. She had only had ascites for 1 month, and her superior mesenteric vein was very small at the time of surgery. This may have contributed to the shunt failure. The other four patients had angiographically demonstrated patent shunts at discharge from the hospital (Fig. 7). One patient (TZ) returned 2 months following surgery and angiography and laparotomy demonstrated a completely thrombosed portal system. Portal vein thrombosis has been reported in other patients with the Budd-Chiari syndrome. One patient (SN) returned four months following surgery with a thrombosed shunt, which was replaced successfully. In examining the thrombosed shunt at operation, it appeared as if the thrombus originated at the atrial anastomosis. Another patient redeveloped ascites 10 months after her shunt, secondary to graft compression by the xiphoid and sternum. The graft was replaced with a prosthesis containing a silastic cuff to avoid compression, and she is free of ascites 5 months later. The final patient maintained graft patency long term and was free of ascites and bleeding at the time of her demise 5 years later.

Because patients with the Budd-Chiari syndrome have a demonstrated tendency for venous thrombosis, anticoagulation following a successful portasystemic shunt, particularly when performed with a prosthesis, has appeal. We have administered long-term coumadin in four patients (DB, JO, SN, TZ) and aspirin and Persantine® in three patients (JO, SN, TZ). However, two patients treated successfully (AL, CS) with the longest follow-up have not received anticoagulants or antiplatelet drugs. The role of these drugs in managing patients with the Budd-Chiari syndrome following successful mesocaval or mesoatrial shunt is not known.

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Discussion

PROFESSOR AKE SENNING (Zurich, Switzerland): I have used another approach to this problem, and that consists of resection of the liver tissue around the caval vein, including the stenosed liver veins, and draining them directly to the right auricle. I have used this in three cases of Budd-Chiari, three different times, but all of them with suprahepatic hindrance.

The first was a 37-year-old dentist. I have a feeling that dentists have some feeling for experimental surgery. (Slide) He had a caval thrombosis and a portal thrombosis and severe ascites, and was 100% an invalid. This is the thrombosed cava; you see the thrombosed and partly recanalized cava.

(Slide) Percutaneous phlebography of the liver veins showed severe stenosis, the liver veins running off to a subphrenic vein to the hemiazygos vein.

(Slide) The next patient was a 26-year-old woman. Retrograde phlebography showed severely stenosed hepatic vein with a runoff partly to a subphrenic vein and partly retrograde to the portal vein.

(Slide) The third patient was a 33-year-old woman with ascites, and in very bad condition. Arteriography showed small vessel disease with a retrograde runoff to the portal vein.

(Slide) The operation consists of a midline incision. We split the diaphragm and the pericardium, and in extracorporeal circulation we place a clamp on the right auricle; then the cava is opened, the web is resected, and we have resected the liver area around the cava here. Then the auricle is sutured around the resected area to the liver capsule.

(Slide) Postoperative percutaneous phlebography shows a very rapid runoff to the resected area of the liver and to the right auricle.

2-½, 1-½, and almost 1 year after surgery, all patients are symptom free and working 100%. The interesting thing is the good result in the patient with small vessel disease. The reason we had to operate on her was massive bleeding from esophageal varices. After the operation, she has had no more bleeding, and one reason is that there is partially an intrahepatic portacaval shunting.

DR. WILLIAM J. MILLIKAN (Atlanta, Georgia):

(Slide) We agree with Dr. Cameron that in patients that have Budd-Chiari syndrome and an infrahepatic obstructed vena cava, mesoatrial shunt is the first in what has become at Emory a two-stage approach. Because of the very high occlusion rate in our hands and elsewhere of meso-caval interposition shunts, and because of our personal experience with obstruction of a mesoatrial shunt, we now suggest that an elective portacaval shunt be performed at a second setting, under the protective umbrella of a mesoatrial shunt, after hepatic decongestion has occurred and intrahepatic vena cava pressure has returned toward normal.

This approach is exemplified in a patient (slide); this 33-year-old female was referred to Emory after having Budd-Chiari's syndrome diagnosed and initially treated with an interposition mesocaval shunt at another hospital. The shunt occluded, and the patient was referred with massive ascites and hepatic dysfunction.

This slide was originally interpreted as having vena caval obstruction, but was then shown not to be clotted, but just obstructed. In other words, this vena cava is patent, and we maintain that this is frequently a problem of interpretation of vena cavograms in patients with Budd-Chiari.

The patient then underwent an urgent mesoatrial shunt. She was rehospitalized 4 months later for elective portacaval shunt. (Slide) This shows the inferior vena cavogram on the same patient 4 months after mesoatrial shunt. The inferior vena caval pressure prior to mesoatrial shunt was 24 mmHg. This has now decreased to 10 mmHg, compared to the right atrial pressure of 6 mmHg.

The second thing which was brought up by Dr. Cameron, which we think is crucial because of its unknown pathophysiology is persistance of hepatic venous occlusion and portal hypertension. A hepatic wedge venogram was done in this patient, and at surgery, where she underwent an elective type of portacaval shunt; the pressure in the portal vein with the mesoatrial shunt occluded was 22 mmHg. Drs. Fulenwider and Henderson then constructed a side-to-side portacaval shunt in this patient and interrupted the mesoatrial shunt.

(Slide) Dr. Warren was the first to suggest this two-stage approach because of the 50% occlusion rate of mesocaval shunts. The mesoatrial shunt reduced hepatic congestion and allows the inferior vena cava pressure to normalize, followed later—4 months in this particular case—with an elective portacaval shunt, after documentation of vena caval obstruction has been relieved.

DR. HARRY H. LEVEEN (Charleston, South Carolina): I have seen 12 patients with this disease, and have received abstracts, after a letter sent to about 500 surgeons, of about 25 other cases with various forms of therapy. I would say that less than 50% of all of these patients would have been suitable for a portacaval or mesocaval type shunt.

One patient had an automobile accident with a New York City taxi, suffered a lacerated liver, and developed a granuloma of the apex of her liver, which produced Budd-Chiari syndrome. She became rehabilitated with a peritoneal venous shunt, and the studies done about 2 years after showed that she had a return of her normal situation.

I believe there must be—and I think it has been the experience of many—patients who have evanescent occlusion recanalized, and perhaps one should be less enthusiastic about rushing in to do some sort of portacaval shunt.

My only experience with atriocaval shunts was in a doctor's wife who had been on the pill, who developed thrombosis of her inferior vena cava, had an atriocaval shunt done elsewhere, and this shunt then occluded; but the unfortunate thing is, when these shunts occlude, and the patient gets severe ascites, the patient has very incapacitating hydrothorax, because of the incision in the diaphragm. And I would just have a word of caution about this type of case, because of this serious complication.

One patient I saw was an Egyptian national, who was sent to me by the Egyptian government having had an inferior vena caval obstruction, and a peritoneal venous shunt had been placed in the patient. This shunt had induced thrombosis of the superior vena cava. Any time we see a patient who has thrombosed a major vessel, and we do a peritoneal venous shunt, we automatically heparinize that patient. It is an indication for heparin. We were amazed to find that there was really no cava at all, and all of the blood was getting back to the right heart through an azygos vein.

(Slide) This shows a picture of this man when he entered. And since there was no available vein, we put this man's sternum and connected his two valves—we put in two valves, with a Y-tube, connected it, and put it directly into the right atrium. (Slide) Here you see a picture of him postoperatively. (Slide) Here is a close-up, showing his sternal