Iliac-Mesenteric-Atrial Shunt Procedure for Budd-Chiari Syndrome Complicated by Inferior Vena Caval Thrombosis

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A 33-year-old woman had a diagnosis of idiopathic Budd-Chiari syndrome complicated by inferior vena caval occlusion. Conservative medical therapy failed to control the symptoms of both portal hypertension and vena caval stasis. Therefore, a prosthetic shunt was placed from the right common iliac vein to the right atrium with a side-arm to the superior mesenteric vein. She exhibited almost complete relief of symptoms and the graft was documented to be patent two weeks postoperatively. In many instances aggressive surgical therapy may help these patients who, in the past, would have been relegated to symptomatic therapy.

BUDD-CHIARI SYNDROME RESULTS from interference with the venous outflow from the liver and can exist in the presence of either intrahepatic or lower inferior vena caval obstruction. Side-to-side portacaval shunts have given successful results in patients with isolated hepatic venous thrombosis who have normal inferior vena cavas and portal veins. When Budd-Chiari syndrome is complicated by inferior vena caval occlusion either in its entire length, in its intrahepatic segment, or in its inferior portion, a standard side-to-side portacaval shunt will not provide benefit or even be feasible. Less common and more complex shunt operations have been devised for this situation, including splenoazygos, azygocaval, splenoatrial, splenopulmonary artery, mesenteric atrial shunts, and others. But to our knowledge, there has never been a report of a combined common iliac vein and superior mesenteric vein-right atrial shunt as reported here in the successful treatment of a Budd-Chiari syndrome, when the inferior vena cava was occluded and therefore not capable of receiving a shunt.

Case Report

In March of 1977 a 33-year-old Greek woman was transferred from her native country to the Ochsner Foundation Hospital for evaluation and treatment of inferior vena caval thrombosis. She had been healthy until six years before admission when there was a sudden onset of increasing abdominal girth without pain, fever, nausea, or vomiting and a bilateral, severe swelling of the legs. She did not have hematemesis, hematochezia, jaundice, or evidence of bleeding. She had no history of hepatitis, blood transfusion, ethynol intake, or cigarette smoking, and she was not taking oral contraceptives or hormones. She had been treated symptomatically with spironalactone (Aldactone) and furosemide (Lasix) and had undergone therapeutic paracentesis and liver biopsies. Treatment of the ascites was mildly successful, but the leg edema persisted. Therefore, one month before admission to this hospital, she had been evaluated with arteriograms, phlebograms, and repeat liver biopsies which were said to be consistent with hepatic vein outflow obstruction and inferior vena cava thrombosis. Other history and family history were not contributory.

On admission the patient was alert, and in no apparent distress. The blood pressure was 110/80 mm Hg; the pulse was 80 beats/min and regular. She was afebrile. The abdomen was distended with ascitic fluid and an umbilical hernia was present. There were many, large dilated veins across the lateral aspects of the abdomen and striae were present inferiorly. The liver was percussible and palpable 8 cm below the subcostal margin in the midclavicular line, but there was no tenderness. Bowel sounds were present. The spleen was palpable and there were no bruits. There was 4+ pitting edema in both legs extending up to the inguinal area with no cyanosis or clubbing. Pulses in both feet were good at both the dorsalis pedis and posterior tibial arteries. She exhibited no signs of portasystemic encephalopathy nor other neurologic findings. The initial impression was that there had been an acute inferior vena caval thrombosis six years in the past and that hepatic vein outflow block had developed, and represented Budd-Chiari syndrome with inferior vena caval obstruction.

The hematocrit was 38%; hemoglobin, 11.9 g/100 ml; WBCs, 7.9, and erythrocyte sedimentation rate 10 mm/hr (Westergren). The platelet count was 461,000/cu mm. Prothrombin and partial thromboplastin time were unremarkable. A 12 factor automated chemical analysis revealed a cholesterol of 134 mg/100 ml; albumin of 3.4 g/100 ml; alkaline phosphatase, 253 mU/ml; LDH, 288 mU/ml; normal SGOT, and a total bilirubin of 2.2 mg/100 ml. The creatinine was 0.8 mg/100 ml, and the electrolytes were normal. SGPT was 17, BSP retention was 26% at 45 minutes. Hemoglobin electrophoresis revealed normal adult hemoglobin. A chest roentgenogram was within normal limits with a prominent azygos vein. Lipid profile was within normal limits. Arterial blood gas determination revealed mild respiratory alkalosis. Urinalysis was normal.

Inferior venacavography revealed near total occlusion at the origin of the inferior vena cava with massive collateralization on both sides (Figs. 1 and 2). Selective celiac arteriography showed massive hepatosplenomegaly and the venous phase showed patent

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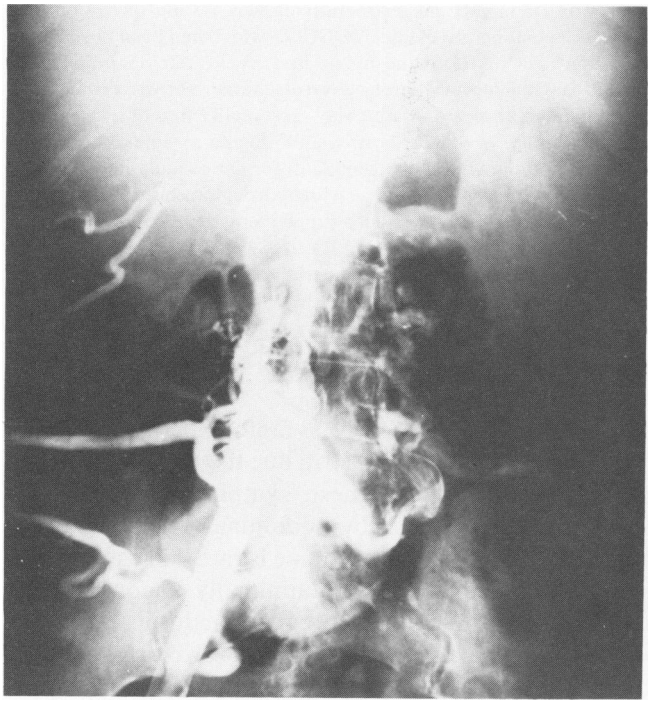


FIG. 1. Occluded inferior vena cava with paravertebral and retroperitoneal collateralization.

splenoportal veins (Fig. 3). Normal kidneys were seen on the aortic flush phase as well as patent right and left hepatic arteries.

A liver scan revealed irregular, patchy distribution in both lobes with shunting to both marrow and spleen and a greatly enlarged spleen. Superior venacavography revealed a right atrial pressure of 4 mmHg, and no hepatic veins could be identified. The inferior vena

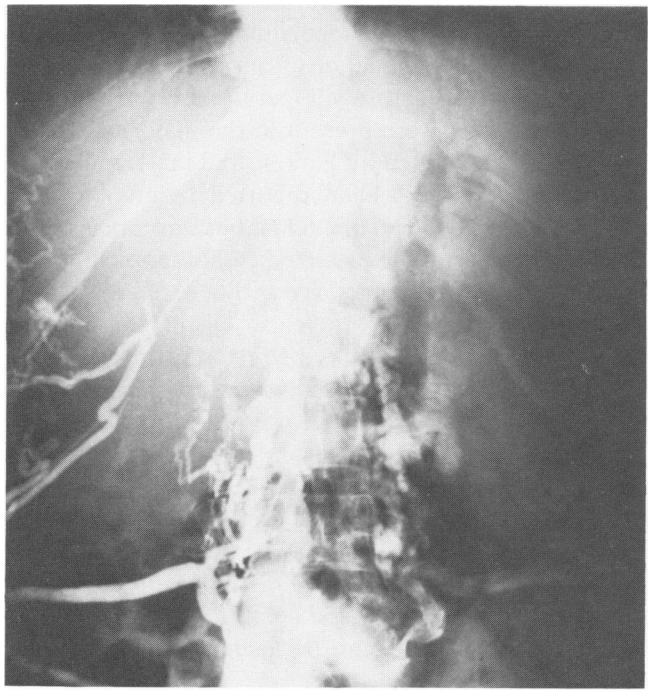


FIG. 2. Later venous phase of Figure 1.

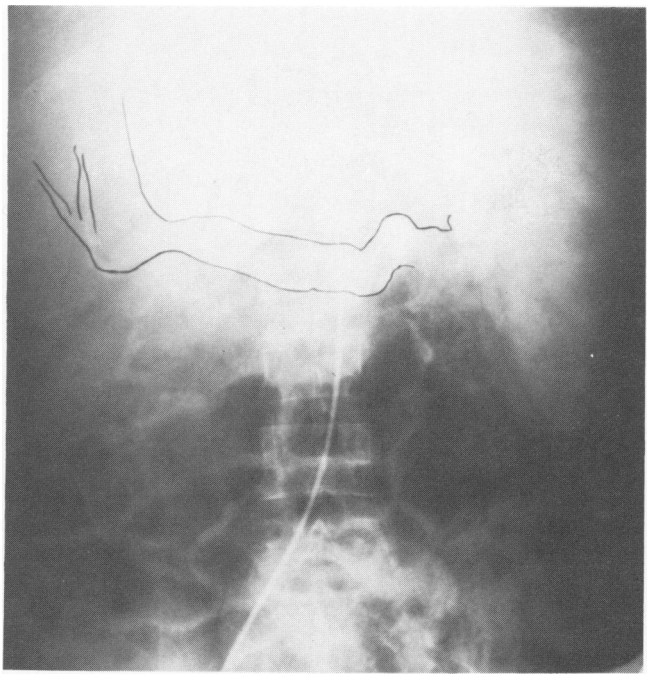


FIG. 3. Venous phase of celiac arteriogram showing normal splenic/portal vein.

cava was occluded at the level of the right renal vein and was filling from it (Fig. 4); the superior vena cava and the azygos vein were greatly dilated.

A needle biopsy of the liver revealed normal appearing hepatocytes and lobules (other than dilatation of central veins) and minimal portal reaction and scarring. The diagnosis was an occluded inferior vena cava from its origin to the right renal vein, and hepatic vein thrombosis, with portal hypertension, chronic ascites, and lower extremity venous stasis. There was no evidence of either active or chronic liver disease.

Because of the severity of the symptoms, the surgical creation of a shunt was indicated. On March 13, 1977 a woven Teflon® graft, 16 mm

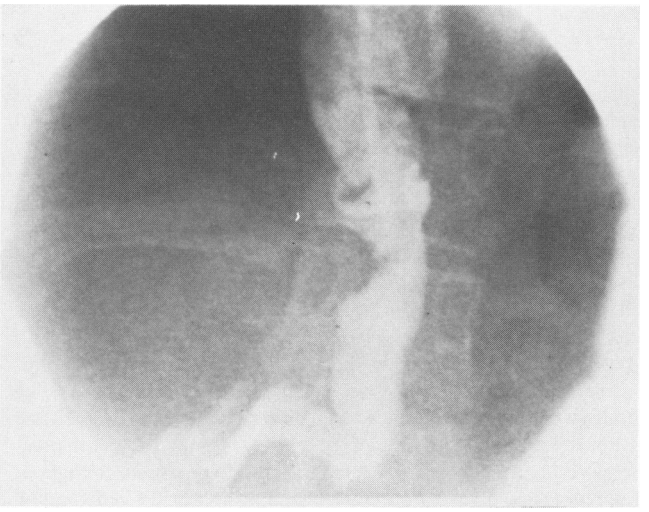


FIG. 4. Intra- and infrahepatic inferior venacavogram from superior approach demonstrating absence of hepatic venous inflow and visualization of right renal vein.

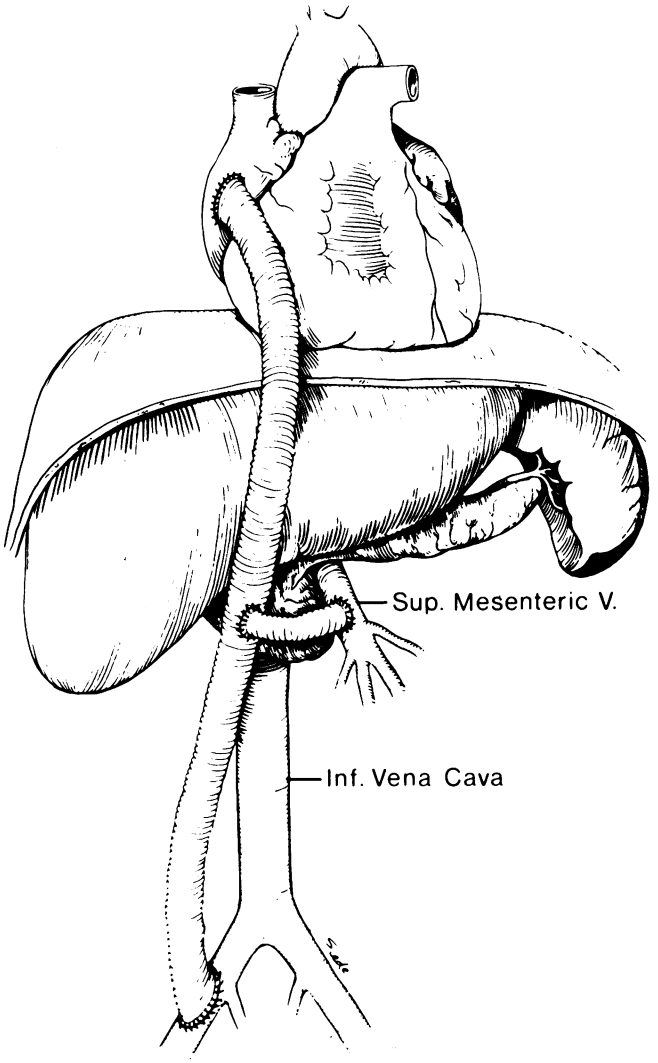


FIG. 5. 16 mm Teflon conduit interposed between the right common iliac vein and the right atrium with 10 mm Teflon side-arm to superior mesenteric vein.

in diameter, was placed end-to-side to the right common iliac vein which was found in an edematous and thickened retroperitoneal space. The graft was passed superiorly, and because of the greatly enlarged liver, it had to be directed anteriorly into the mediastinum through the space left vacant by excision of the xyphoid process of the sternum, and anastomosed to the right atrium. A 10 mm side-arm graft, 4 cm long, was placed in an H fashion from the superior mesenteric vein to the initial graft (Fig. 5). Heparin was given for anticoagulation during the installation of the vascular prosthesis. She required eight units of whole blood during the procedure.

Postoperatively, the patient did well. She was treated with anticoagulants and diuretics, and her weight decreased from 72 to 60 kg within a week of surgery. The ascites almost completely resolved and the lower extremity edema resolved entirely. On the thirteenth postoperative day a phlebogram showed the graft to be patent throughout its length (Figs. 6a and b). Doppler studies confirmed venous flow through the prosthesis from abdomen to mediastinum. Throughout the postoperative course the patient's renal functions were normal and her total protein remained within normal limits. The bilirubin which was 2.2 mg/100 ml preoperatively was 1.3 mg/100 ml on the nineteenth postoperative day. The SGOT which had been normal on admission rose to 70 mU/ml in the first week after operation and then fell to within normal limits before discharge. She was discharged on the nineteenth postoperative day, essentially free of ascites and lower extremity swelling. She was afebrile and asymptomatic. She was given sodium warfarin (Coumadin) for anticoagulation and was to continue taking spironolactone with hydrochlorathiazide (Aldactazide) 25 mg q.i.d. and potassium supplements, and to follow a low salt diet.

At last report, four months postoperatively, she was doing well and has continued to be followed-up by her physicians in Greece.

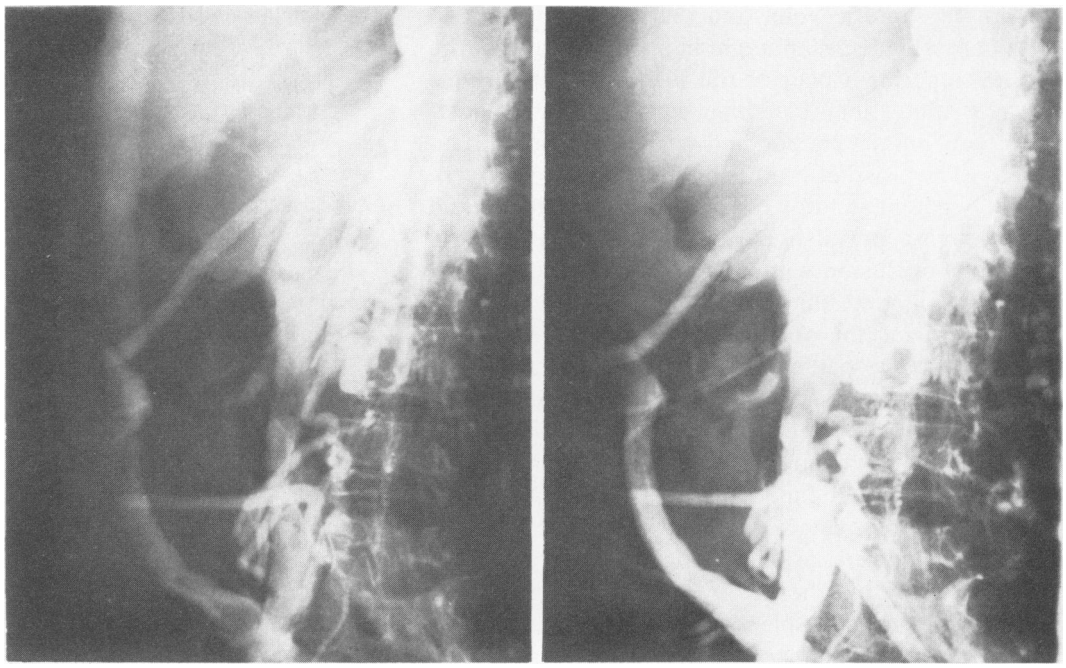
Discussion

The most common early symptom of Budd-Chiari syndrome is an increasing girth due to gross ascites and usually hepatomegaly. These symptoms may be accompanied or preceded by abdominal pain or the patient may be pain-free.[13] Jaundice is rare and usually the liver function tests are only minimally abnormal. On initial physical examination caput medusae is rarely noticed, but pitting edema of the lower extremities or the trunk has been noticed in slightly over 50% of patients.

The causes of Budd-Chiari syndrome are many and include polycythemia rubra vera, tumors in and about the liver, trauma, leukemia, sickle cell anemia, hydatid diseases, exogenous toxins, paroxysmal nocturnal hemoglobinuria, and congenital caval obstructions, as well as, a growing incidence related to oral contraceptive use.[1-4,6-8,13,18]

In Parker's classic study of 1959[13] 30% of the cases had certain, or reasonably certain, causes for Budd-Chiari syndrome, but the remaining 70% were idiopathic, and even in publications from more recent years the majority of cases are idiopathic. Lesions in the inferior vena cava were present in the majority of the idiopathic cases studied by Parker, and in a number of cases, thrombosis has been reported to involve the entire vena cava and hepatic veins, but thrombosis has also been reported to be localized from the renal veins to the diaphragm and it may or may not extend into the hepatic veins and their radicals. Occlusion of the hepatic vein ostia alone has been found in approximately 40% of all the idiopathic cases. Thrombi of different ages are nearly always present in the acute and subacute stages of the disease, and even in the majority of clinically acute cases, completely fibrosed lesions can be found. Thus, in all but the final stages of the process, lesions of different hepatic veins are usually of widely varying age as judged histologically, and are located at various levels of the hepatic venous outflow. Portal vein thrombosis can be seen in 20% of these cases.

Diagnostic aids include radiocolloid liver scans,



FIGS. 6A. Oblique view of postoperative phlebogram demonstrating graft patency. 6B. Later injection phase.

which usually show central localization of the colloid. The central localization is explained by the compensatory hypertrophy of the caudate lobe because of its more efficient perfusion and outflow which is usually not destroyed in primary hepatic vein thrombosis.[19] The narrowing and distortion of the inferior vena cava due to compression by the caudate lobe will often give a typical pencil-pointing of the intrahepatic and inferior vena cava in cases uncomplicated by inferior vena caval thrombosis.

Definitive preoperative diagnosis may require both inferior and superior venacavography, as well as abdominal and celiac axis arteriography, since other diseases such as right-sided heart failure, constrictive pericarditis, membranous obliteration of the inferior vena cava, and tumor thrombus in hepatic veins or inferior vena cava may simulate Budd-Chiari syndrome.[20] Japanese authors emphasize that simultaneous superior and inferior venacavography is necessary if one is not to miss the inferior vena caval membranes which are said to be the most common cause of Budd-Chairi syndrome among the Japanese population.[6-8,18] The celiac arteriogram is a necessary preoperative study in that any contemplation of shunting procedure requires demonstration of the anatomy as well as the adequacy of the hepatic arterial inflow. The splenoportogram done indirectly from the celiac arteriogram will reveal the presence and adequacy of the contributories to the portal vein which is necessary knowledge especially when shunts other than side-to-side are contemplated. Obviously, right heart catheterization may be useful and an attempt is usually made to cannulate the hepatic vein ostia themselves, which may or may not be possible even with a clinical picture consistent with complete hepatic vein thrombosis. In many cases occlusion of the hepatic veins does not occur at the ostia, but in more proximal, medium, hepatic vein channels such that the ostia themselves may appear normal radiologically and may even give a reading for hepatic wedge and free hepatic vein pressure measurements.

The natural history of the disease dictates that, when a patient fails to improve on adequate medical therapy, a decompressive shunt is the only chance for relief of symptoms and protection from the effects of chronic portal hypertension. Langer's review of the results of treatment in 55 cases published since 1959 emphasized the importance of definitive surgical therapy and recommended it for all but the most rapidy resolving cases.[9] Five of 14 patients given definitive operations died, but the other 9 improved; whereas 29 of 41 medically treated patients died, and only 7 improved.[9]

Most of the reported patients successfully treated with shunts are those whose disease was not complicated by concomitant inferior vena caval obstruction. Side-to-side portocaval or mesocaval shunts have been relatively successful in these less complicated situations.[16,14]

For patients who have concomitant inferior vena caval obstruction more complex and less successful shunt operations have been devised. Hales and Scatliff[5] in 1966 treated a man with chronic Budd-Chiari syndrome and inferior vena caval obliteration with a Dacron® Y-graft placed between the right atrium

superior mesenteric vein, and the right internal iliac *artery*, crossing the diaphragm in a trough through the anterior superior aspect of the left lobe of the liver. The gastrointestinal bleeding was eliminated postoperatively but the patient died on the tenth postoperative day. The prosthetic graft and the anastomoses were all widely patent at autopsy. Sen et al.[18] in 1967 treated a patient with portal hypertension and suprahepatic vena caval occlusion with a Dacron shunt from portal vein to the right atrium; however, the patient died two weeks following the operation. At autopsy the shunt was found to be patent. Lataste and Albou in 1971 reported an unsuccessful attempt at splenomesenteric-atrial bypass.[10] Leger et al. in 1966 utilized a Teflon prosthetic implant from the splenic vein to the left pulmonary artery which failed, and in another patient, attempted a splenic vein to right atrial shunt.[11] O'Hara et al. in 1963 were probably the first to report the cavoatrial shunt but their results had not been good.[12] Both azygocaval and splenoazygos shunts have been used with occasional success.[21,15] However, to our knowledge a shunt from the common iliac vein to the right atrium with a side branch to the superior mesenteric vein has never before been reported. Such a shunt was necessary for our patient to alleviate the symptoms of vena caval occlusion. The patient's onset and clinical course were characteristic of the Budd-Chiari syndrome and the thrombosis of the inferior vena cava was certainly a related event. The objective and subjective improvement was dramatic and should encourage more optimism in the surgical treatment of such patients.

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