Protean Manifestations of Pylethrombosis

A Review of Thirty-Four Patients

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Thirty-four adult patients with portomesenteric venous occlusion (PVO) were reviewed. In 11 with hepatic cirrhosis, PVO was usually heralded by worsening ascites often with varix hemorrhage; mortality was high. Four with isolated portal block had varix hemorrhage without ascites. All of these patients survived despite recurrent hematemesis when portal decompression was not feasible in two patients. Eight others (5 agnogenic and 3 with hypercoagulability), experienced sudden abdominal pain with a clot typically propagated into mesenteric tributaries with ileojejunal infarction; survival was related to the promptness of operation and the extent of bowel ischemia. Of five patients with intraabdominal sepsis and pylephlebitis, only one survived. In the final six patients, PVO occurred with intraabdominal carcinoma. Five had progressive ascites, cachexia, and an early death. Imaging techniques included plain and contrast roentgenograms, ultrasonography, and for definitive diagnosis direct portography (operative or splenoportogram), indirect portography (splanchnic arteriovenogram), and computed tomography. Thirteen of 34 patients had ascites, and in nine of 11 patients examined, protein concentration of ascitic fluid was extremely low (< 0.6 g/dl). Clinical presentation of PVO varies, depending on acuteness and extent of visceral venous blockade, severity of portal hypertension, auxiliary venous collateralization, and regional lymph flow. Inciting factors include endothelial damage and blood hypercoagulability from trauma, infection, stagnant circulation, blood dyscrasia, and malignancy. Improved imaging now allows early diagnosis.

PORTOMESENTERIC VENOUS THROMBOSIS is usually associated with syndromes of visceral ischemia and portal hypertension. Although bowel infarction and death may follow, as in occlusive and nonocclusive mesenteric arterial insufficiency, symptoms often have a long prodrome, simulate severe gastroenteritis, and vary depending on extent and suddenness of venous blockade.[1] Thus, ascites, varix hemorrhage, and bowel infarction, alone or together, occur after pylethrombosis much as edema, venous varicosities, and impending gangrene (phlegmasia cerulea dolens) follow deep vein iliofemoral thrombosis. Because imaging of the portal venous system is vastly improved and pylethrombosis is, therefore, potentially amenable to accurate preoperative and antemortem diagnosis, we examined our cumulative clinical experience with portal vein occlusion (PVO). Specifically excluded were patients with strangulated bowel from volvulus, intussusception, or other closed loop obstruction as well as those where PVO developed after abdominal operations such as splenectomy or distal splenorenal shunt.

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Clinical Material

Thirty-four patients aged 15 to 75 years with documented (radiologic imaging, autopsy, laparotomy) portal venous occlusion form the basis for this report. The major clinical features, including treatment, are summarized in Table 1.

Hepatic Cirrhosis

Patients 1 through 11 (Table 1). The largest group of patients with pylethrombosis had coexistent hepatic cirrhosis. Of 11 patients, nine had conditions related to alcoholism (patients 1, 3, 5–11); the others had conditions related to amethopterin (methotrexate) treatment for refractory psoriasis (patient 2) and viral hepatitis (patient 4). Development of PVO was usually heralded by the sudden appearance or worsening of ascites, explosive hemorrhage from esophagogastric varices, or both. In two patients, PVO was precipitated by tumor invasion from superimposed primary liver cell carcinoma. In another two, hemorrhagic small bowel infarction was the major manifestation, limited to a jejunal segment in one but extensive in the other. Overall, occurrence of PVO with cirrhosis was highly lethal with eight of 11 patients dying soon after appearance or exacerbation of

TABLE 1. *Clinical Features of Pylethrombosis*

Patient	Age/Sex	Etiology	Symptom(s)	Finding(s)	Definitive Diagnosis	Treatment	Outcome
1	40/M	Cirrhosis	Hematemesis	Varix hemorrhage	Splenoportogram, laparotomy	Proximal S-R shunt	Survived†
2	65/F	Cirrhosis	↑ abd girth	Ascites	Splenoportogram, autopsy	Portacaval shunt	Dead
3	65/M	Cirrhosis	↑ abd girth; hematemesis	Ascites; varix hemorrhage	Arterioportogram, autopsy	P-J shunt, sclerotherapy	Dead
4	70/M	Cirrhosis*	↑ abd girth; hematemesis	Ascites; varix hemorrhage	Splenoportogram, autopsy	None	Dead
5	50/M	Cirrhosis*	↑ abd girth	Ascites; abd mass	Arterioportogram	None	Dead
6	55/M	Cirrhosis	↑ abd girth; hematemesis	Ascites; varix hemorrhage	Laparotomy	M-C shunt	Dead
7	58/F	Cirrhosis	↑ abd girth	Ascites; SBP; uremia	Autopsy	Antibiotics, diuretics	Dead
8	61/M	Cirrhosis	↑ abd pain; diarrhea	Segmental jejunal infarction	Laparotomy	Bowel resection	Survived
9	47/M	Cirrhosis	↑ abd pain; diarrhea	Ileojejunal infarction	Laparotomy	Bowel resection	Dead
10	55/M	Cirrhosis	↑ abd girth	Ascites	D-S arterioportogram	P-J shunt	Alive 3 yrs
11	75/M	Cirrhosis	↑ abd girth; hematemesis	Ascites; varix hemorrhage	Splenoportogram	P-S shunt	Dead
12	33/F	None	Hematemesis	Varix hemorrhage	Arterioportogram	Distal S-R shunt	Alive 2 yrs
13	20/F	None	Hematemesis	Varix hemorrhage	Operative portogram	Laparotomy	Alive 1 yr
14	25/M	None	Hematemesis	Varix hemorrhage	Splenoportogram	Proximal S-R shunt	Alive 5 yrs
15	15/M	None	Hematemesis	Varix hemorrhage	Operative portogram	Makeshift P-S shunt (failed)	Alive 12 yrs
16	55/M	None	Abd pain, anorexia	Splenomegaly; bowel edema	Arterioportogram, laparotomy	Laparotomy	Alive 1 yr
17	29/M	"Crash-diet"	Abd pain, anorexia, diarrhea	Ileojejunal infarction	Laparotomy; arterioportogram	Bowel resection	Alive 3 yrs
18	32/M	None	Abd pain, anorexia, diarrhea	Segmental jejunal infarction	Laparotomy; arterioportogram	Bowel resection	Alive 4 yrs
19	25/M	Ovulen	Abd pain, anorexia, diarrhea	Ileojejunal infarction	Operative portogram	Bowel resection	Dead
20	71/F	None	Abd pain, anorexia, diarrhea	Segmental jejunal infarction	Laparotomy	Bowel resection	Survived
21	62/M	None	Abd pain, anorexia, diarrhea	Segmental jejunal infarction	Laparotomy	Bowel resection	Survived
22	71/F	None	Abd pain, anorexia, ↑ abd girth	Ascites, bowel ischemia	Arterioportogram; autopsy	Nonoperative	Survived‡
23	63/F	P. vera	Abd pain, anorexia	Ileojejunal infarction	Autopsy	Nonoperative	Dead
24	37/M	Appendicitis	Abd pain; hypotension	Ileojejunal infarction	Autopsy	Bowel resection	Dead
25	70/M	Diverticulitis	Abd pain, chills, fever	Perforated sigmoid colon	Autopsy	Antibiotics	Dead
26	55/M	Pancreatitis	Abd pain, vomiting	Necrotizing pancreatitis	CT; arterioportogram	Nonoperative	Dead
27	67/F	Amebiasis	Abd pain, chills, fever	Amebic colitis; hepatitis	Autopsy	Drainage, metronidazole	Dead
28	54/M	Diverticulitis	Ketoacidosis	Liver abscesses	CT; arterioportogram	Drainage	Alive 1 yr
29	59/M	Ca pancreas	Abd pain; weight loss	Abd mass; ascites	Autopsy	Supportive	Dead
30	60/F	Ca pancreas	Abd pain; ↑ abd girth	Ascites	Autopsy	Supportive	Dead
31	50/F	Ca pancreas	↑ abd girth	Ascites	Autopsy	Supportive	Dead
32	65/F	Ca kidney	Abd pain; weight loss	Metastatic hypernephroma	CT; autopsy	Supportive	Dead
33	62/M	Ca stomach	Abd pain	Jejunal infarction	CT	Bowel resection, gastrectomy	Dead
34	68/F	Ca pancreas	↑ abd girth, weight loss	Ascites	Autopsy	Supportive	Dead

S-R, splenorenal; P-J, peritoneojugular; M-C, mesocaval; P-S, portasystemic; SBP, spontaneous bacterial peritonitis; P. vera, polycythemia vera; CT, computed tomography; D-S, digital-subtraction; Ca, carcinoma; and abd, abdominal.

* Also hepatoma.
† Dead 6 years.
‡ Dead 2 years from a perforated ileum.

symptoms. Seven of eight (patients 2–7, 11) died after sudden development or rapid progression of ascites. In five of these eight (patients 3–6, 11), refractory ascites was accompanied by varix hemorrhage, the immediate cause of death.

In patient 7, superimposed bacterial peritonitis (*Escherichia coli*) led to progressive uremia and terminal hepatocellular failure despite administration of massive antibiotic and diuretic drugs. In patient 2, venous thrombosis had propagated throughout both the greater

splanchnic (mesenteroportal) and lesser splanchnic (gastrosplenic) venous systems. A LeVeen shunt was inserted to redistribute extracellular fluid, thus reversing the patient's refractory ascites and renal insufficiency. But subsequent varix hemorrhage proved fatal despite repeated transesophageal sclerotherapy. An autopsy confirmed occlusion of all major visceral venous trunks (Fig. 1). In both patients with hepatoma (patients 4 and 5) a tumor with a clot obliterated the greater splanchnic system, but the gastrosplenic circulation remained patent (Fig. 2). In four patients (patients 1, 2, 6, 11), venous occlusion was limited to the portal vein proper (Fig. 3); outward manifestations were bleeding varices (patient 1), intractable ascites (patient 2), or both (patients 6 and 11). As shown in Table 1, early death was avoided only when portal venous decompression was successful (patient 1), extracellular fluid redistributed in the absence of varix bleeding (patient 10), or bowel infarction was limited and amenable to segmental resection (patient 8). Although patient 1 survived portal decompression, death 6 years later followed an accident in which he was driving while intoxicated.

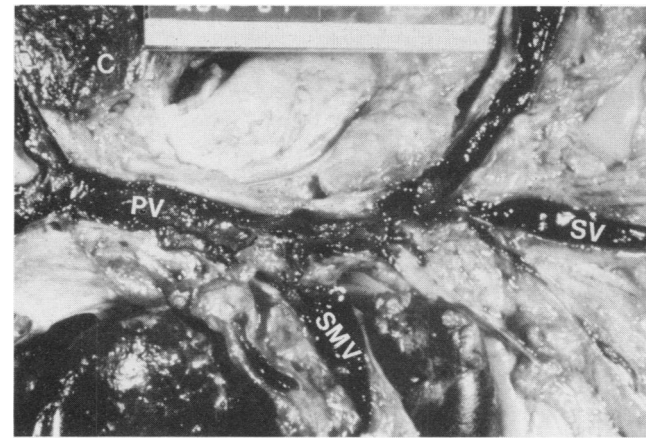


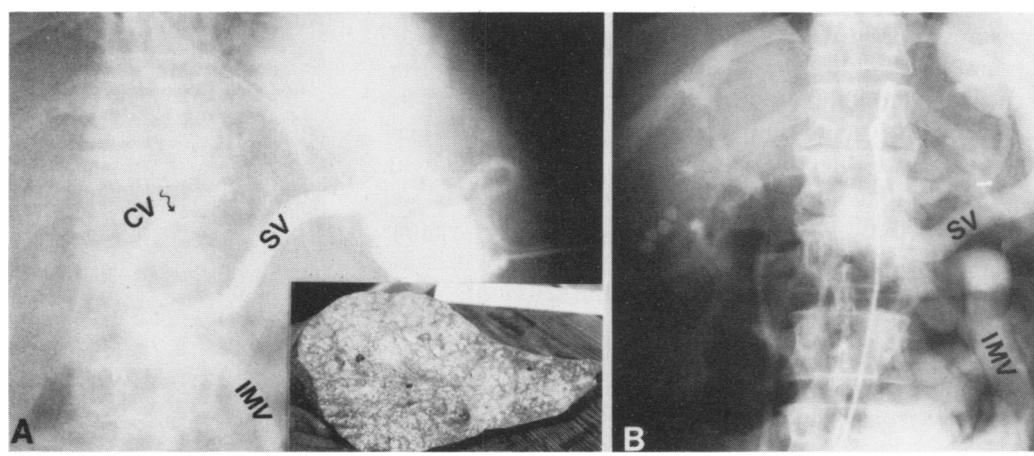
FIG. 1. Diffuse venous thrombosis of the portal system in a 65-year-old man with alcoholic cirrhosis (patient 3). PV, portal vein; SV, splenic vein; SMV, superior mesenteric vein; C, cirrhosis.

Cavernous Transformation

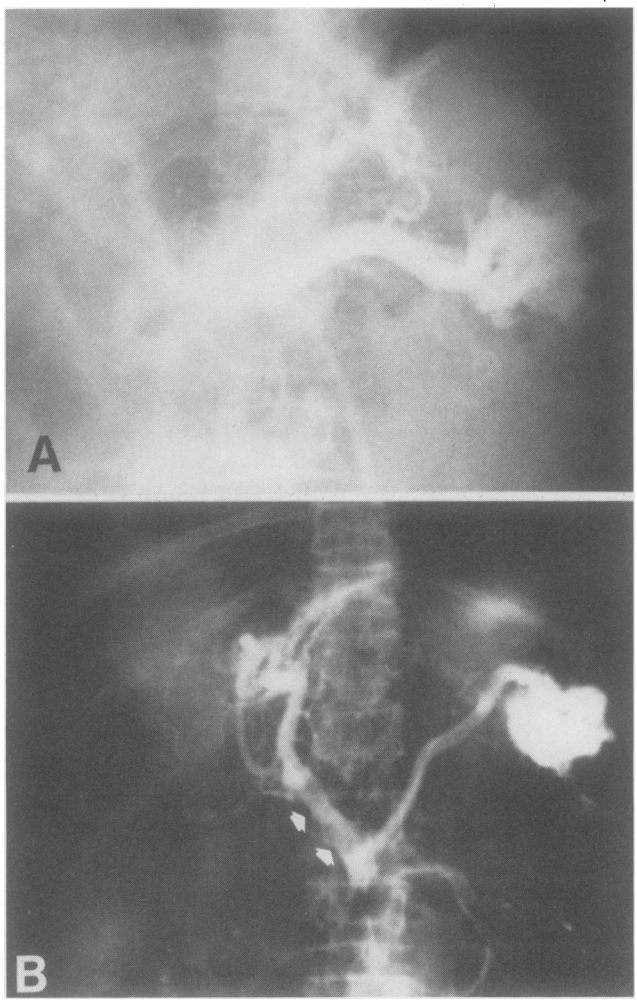
Patients 12 through 15 (Table 1). In four patients, venous occlusion was limited to the portal vein proper, probably existed for a long time before onset of symptoms. In each patient, the initial manifestation was massive hematemesis from rupture of esophagogastric varices. Ascites was notably absent. Preoperative diagnosis was confirmed either by celiac-mesenteric arterioportography (patient 12), splenoportography (patient 14) (Fig. 4), or (patients 13 and 15) by direct intraoperative portography (Fig. 12) *via* either an omental or a jejunal venous tributary (patients 13 and 15). Table 1 shows that each patient is still alive, although, in patients 13 and 15, in whom the portal system was not decompressed, intermittent, self-limited hematemesis had recurred. Despite this untoward sequela, both patients are alive 7 and 10 years later, respectively.

Acute Pylethrombosis

Patients 16 through 23 (Table 1). In eight patients, the onset of symptoms was sudden, characterized by intense epigastric-periumbilical pain, anorexia, and severe diarrhea. Often, the seriousness of the condition was appreciated only after the symptoms had persisted for several days or after they had become complicated by hematochezia or hypotension. Six patients were initially thought to have acute viral gastroenteritis, a diagnosis consistent with abdominal plain roentgenograms showing segmental, slightly dilated loops of small intestine (Fig. 5). Typically, portomesenteric thrombosis was extensive, with the clot propagating into peripheral venous radicles with diffuse ileojejunal hemorrhagic infarction (Fig. 6).



FIGS. 2A and B. Splenoportogram (*A*) and superselective splenic arterioportography (*B*) demonstrating portal vein occlusion in two patients by hepatoma superimposed on hepatic cirrhosis (*inset A*). SV, splenic vein; IMV, inferior mesenteric vein; CV, coronary vein.

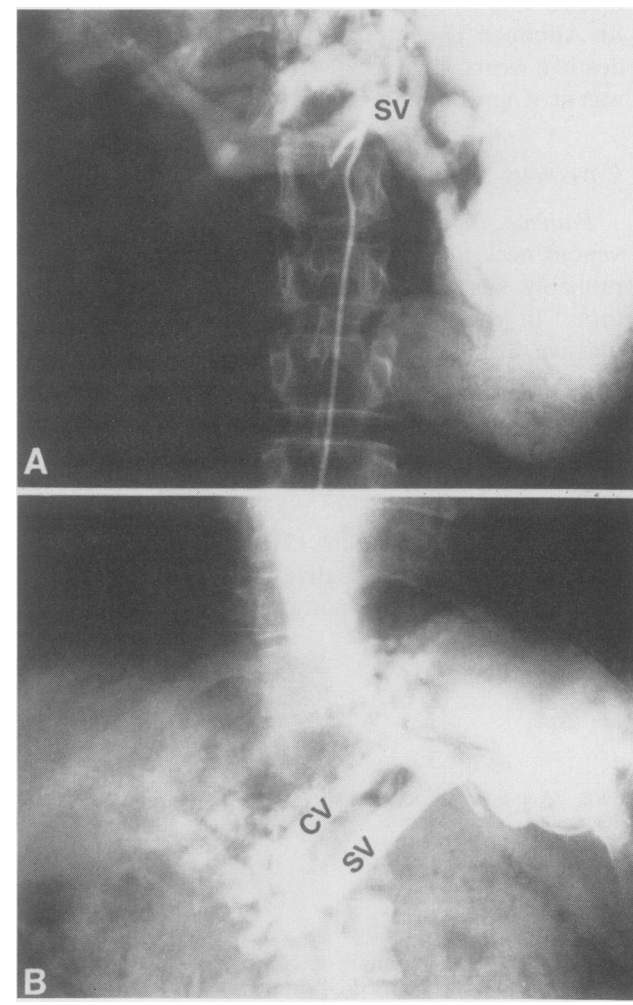


FIGS. 3A and B. Splenoportography in patients 1 and 2, with hepatic cirrhosis and superimposed extrahepatic portal vein occlusion. *A*, recanalization and periportal venous collateralization are extensive; *B*, the portal vein contains thrombi (arrows) and is totally blocked in the porta hepatis.

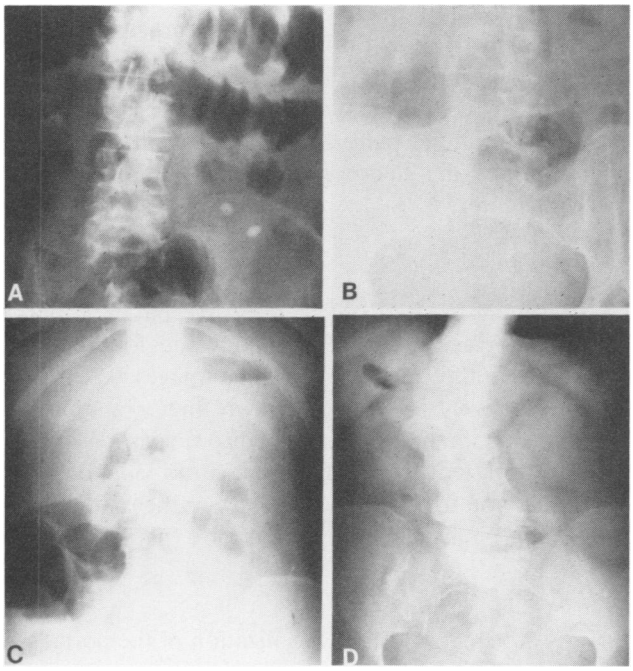
In three patients, the contributing factors were polycythemia vera, oral contraceptive usage (Ovulen®), and severe caloric-intake restriction ("crash-diet" with 60 pound weight loss in 30 days). In the other five, no coexistent condition was incriminated; this justified the diagnosis of acute agnogenic pylethrombosis. Patient survival was directly related to the promptness with which the patient was operated on and the extent of bowel infarction. Despite the diffusiveness of venous occlusion, colonic infarction was rare. Bowel necrosis was generally limited to the ileum and jejunum. When much of the ileum or jejunum was gangrenous, reoperations with further debridement and resection of residual nonviable bowel were common. The clinical course was typically stormy and prolonged even when death was averted. Anticoagulant drugs were used short-term in patients 18 and 19 (*i.e.*, superior mesenteric arterial infusion of heparin), and chronically as coumadin® (#18), but omitted altogether in patients 17 and 22. In patients 17, 18, and 22, antithrombin levels in peripheral blood were normal. As shown in Table 1, prolonged survival depended primarily on the feasibility and success of resecting ischemic bowel. In patient 16, acute symptoms spontaneously abated and laparotomy failed to reveal an explanation for portal vein occlusion. Despite intraoperative findings consistent with portal hypertension, he remains well.

Septic Pylephlebitis

Patients 24 through 28 (Table 1). In five patients, pylethrombosis was associated with other acute intraab-



FIGS. 4A and B. Chronic extrahepatic portal block in two patients without liver disease, demonstrated by celiac arterioportography (*A*, patient 12) and splenoportography (*B*, patient 14). SV, splenic vein; CV, coronary vein.



FIGS. 5A–D. Plain roentgenograms in acute pylethrombosis, demonstrating markedly thickened small bowel wall and valvulae from edema (*A*), segmentally dilated ileojejunal loops with air-fluid levels and lack of colon air (*B* and *C*), and diffuse ground-glass appearance from sequestered intraperitoneal and intraluminal fluid with paucity of gas shadows (*D*).

dominal conditions (Figs. 7 and 8). In three patients, portal thrombophlebitis developed in conjunction with portal pyemia (suppurative appendicitis and sigmoid diverticulitis, both perforated and nonperforated). In one patient, it developed from amebic colitis with intrahepatic abscesses, and in the final patient, the only one to survive, pylethrombosis accompanied acute necrotizing pancreatitis.

Intraabdominal Carcinoma

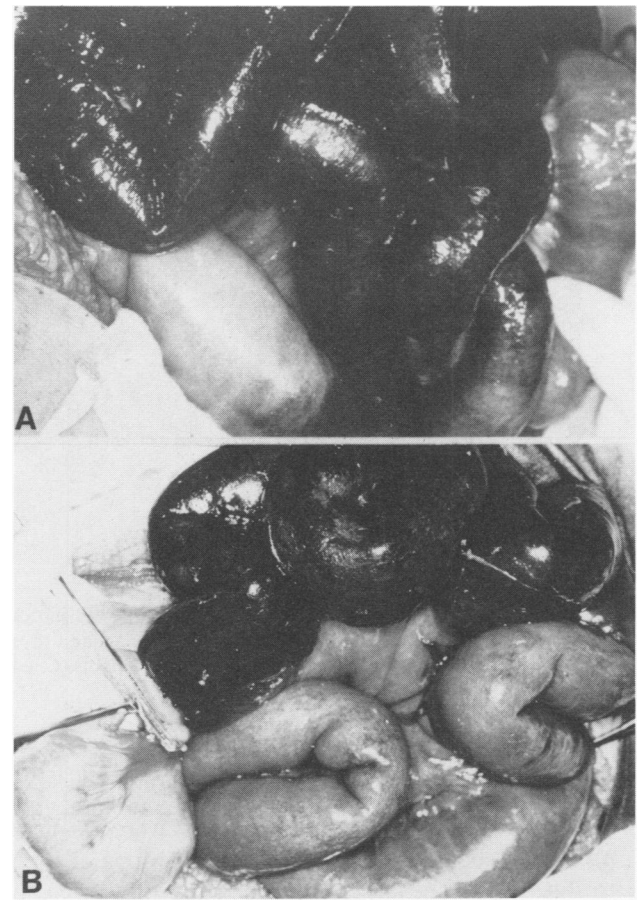
Patients 29 through 34 (Table 1). Pylethrombosis developed in six patients with intraabdominal carcinoma. In four, the primary tumor was in the head of the pancreas and in two others, the right kidney (Fig. 8) and distal stomach (Fig. 9), respectively. Five patients developed progressive ascites, abdominal pain, profound weight loss, and severe anorexia. The sixth had acute bowel infarction. In each, death ensued shortly thereafter. Diagnosis was confirmed at autopsy or operation, although in two recent patients (32 and 33), antemortem diagnosis was made by computed tomography (Figs. 8 and 9).

The primary clinical manifestations of pylethrombosis of divergent origins are summarized in Table 2.

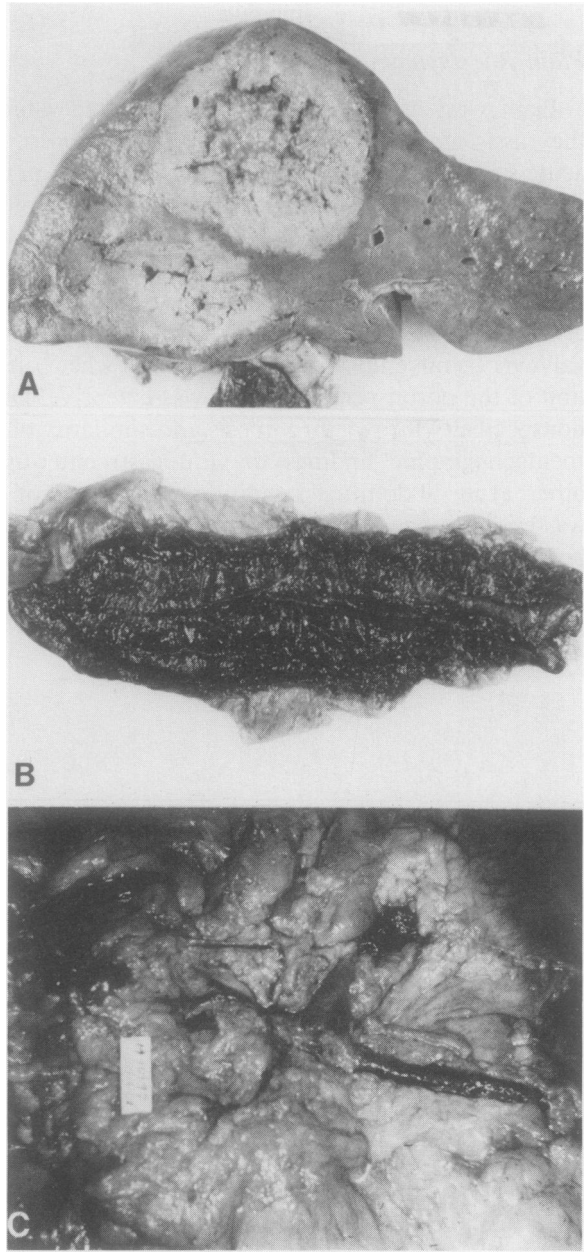
Imaging

Plain Roentgenograms

Plain x-ray abdominal films are of limited value, but they assisted in the diagnosis of pylethrombosis. With acute symptoms, nonspecific segmental dilatation of the small intestine was common often in conjunction with a paucity of bowel air and an overall ground-glass appearance suggestive of sequestered intraperitoneal or intraluminal fluid. Intensive digestive tract edema was also evident by a thickened bowel wall or swollen valvulae connivente (Fig. 5), particularly where obstruction of the portal venous system was extensive. Despite nonspecificity for pylethrombosis and similarity of these roentgenographic findings to viral gastroenteritis and other acute abdominal conditions, suspicion of PVO was raised by the relative lack of colon air despite small bowel distention along with fluid sequestration within both bowel lumen and peritoneal cavity.



FIGS. 6A and B. Diffuse ileojejunal infarction associated with acute pylethrombosis. Despite apparently viable enteric segments (light grey areas), venous clot had visibly propagated throughout the small bowel mesentery in patients 17 (*A*) and 19 (*B*).



FIGS. 7A–C. Septic endophlebitis with thrombosis of the portal system associated with large intrahepatic abscess (*A*), secondary to acute amebic colitis (*B*, patient 27), and suppurative appendicitis (*C*, patient 16).

Contrast GI Series

Performed only where symptoms were vague and prolonged, barium contrast roentgenographs of the gastrointestinal tract largely corroborated other diagnostic maneuvers. Whereas it commonly revealed esophagogastric varices in patients with hepatic cirrhosis and cavernous transformation of the portal vein, the diagnosis had usually been previously established by transesophageal endoscopy or angioportography. In patient 22, small bowel follow-through revealed ischemic changes in the distal ileum (Fig. 10) consistent with known pylethrombosis and symptoms of persistent anorexia and postprandial pain. In this individual, ischemic perforation of the ileum led to peritonitis and death 8 months later.

Ultrasonography

Although carried out in only five patients, real-time ultrasonography was of limited value for diagnosis of portal vein thrombosis, even when images were examined retrospectively in light of known angiographic and autopsy documentation of pylethrombosis. Pylephlebitis was suggested from abnormal intrahepatic channels known not to be dilated biliary radicles by previous negative biliary-hepatic scintigraphy (patient 28).

Angiography

As anticipated, direct visualization of the portal system was the most accurate preoperative or antemortem way to establish the diagnosis of pylethrombosis. In our early experience, splenic pulp puncture with direct portography was definitive (Figs. 2–4), but more recently, celiac-mesenteric arterioportography, often with flow-enhancement by regional infusion of vasorelaxants (papavarine, tolalozine) was preferred (Figs. 2 and 4). On occasion, digital-subtraction computed splanchnic arterioportography was chosen. This was the case particularly when a limited volume of contrast infusion was desired as, for example, with borderline renal function (Fig. 11). Where differential diagnosis was especially difficult, intraoperative portography after cannulation of a portal venous tributary was definitive. Thus, in patients 13 and 15, with chronic extrahepatic portal block and varix hemorrhage where the spleen was absent and visceral arterioportography less than optimal, accurate anatomic delineation was accomplished at laparotomy by direct portography (Figs. 12A and 12B). Similarly, with acute pylethrombosis and bowel ischemia, the diffuseness of propagated venous occlusion was delineated by intraoperative portography (Fig. 12C). Other advantages included corroboration of preoperative indirect portography and of portasystemic shunt patency (Fig. 12D).

Computed Tomography (CT)

Although used only relatively recently, abdominal CT scanning has proven extremely useful. Not only are dilated portal channels with "fuzzy edges" delineated by intravenous contrast infusion consistent with pylethrombosis, but concomitant intraabdominal conditions such as tumors and abscesses are also revealed (Figs. 8–10).

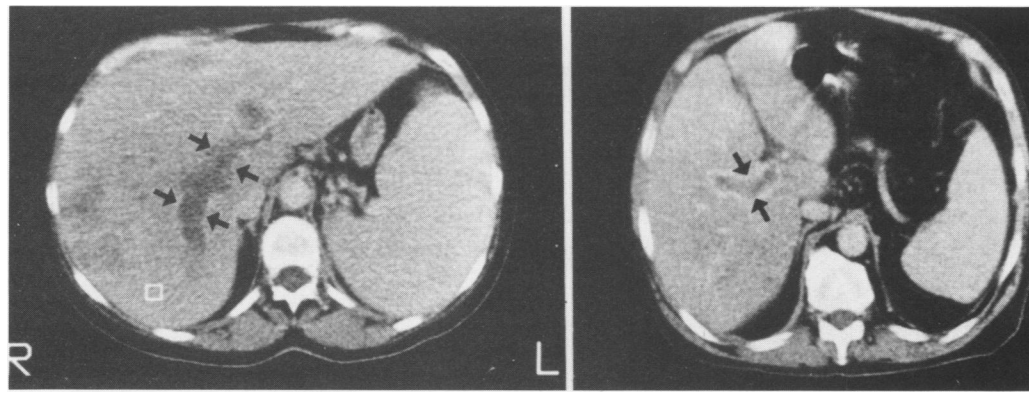
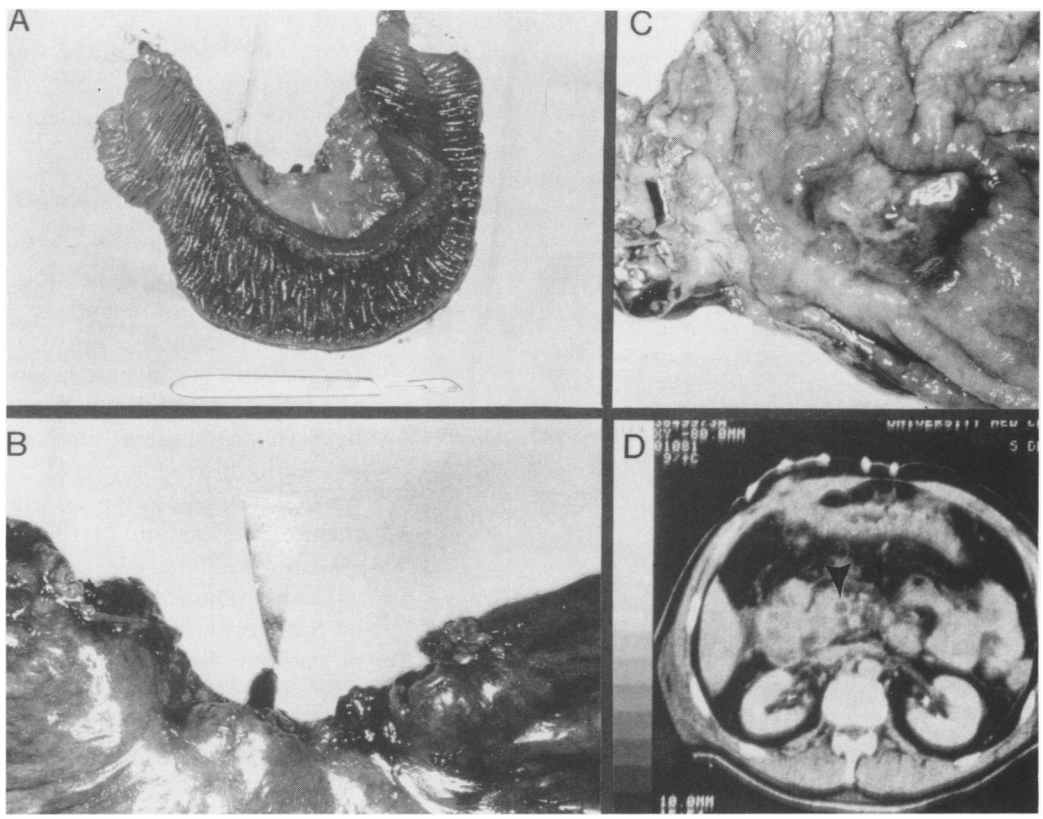


FIG. 8. Abdominal computed tomography with intravenous contrast enhancement demonstrating portal vein thrombosis associated with metastatic renal cell carcinoma (left, patient 32) and intrahepatic pyogenic abscess secondary to perforated sigmoid diverticulitis (right, patient 25). Note the dilated venous channels within the liver (arrows) with contrast outlining only the edges of portal radicles.

Intraabdominal Fluid

Ascites (*i.e.*, grossly palpable intraabdominal fluid) developed in 13 patients with pylethrombosis. Eight had hepatic cirrhosis (patients 2–7, 10, 11); three, carcinoma of the pancreas (patients 29–31); one, hypernephroma (patient 32); and one, diffuse portomesenteric venous thrombosis (patient 22). Although portal hypertension was documented by measurement in only four cases, coexistent esophagogastric varices or varix hemorrhage was demonstrated in five others and intraabdominal portasystemic venous collateralization and/or splenomegaly in the remainder. It is likely, therefore, that portal hypertension existed in all. Of these 13 patients, total protein of the ascitic fluid was measured in 11. In nine the ascitic fluid was extremely low in protein content (*i.e.*, <0.6 g/dl or <10% of plasma) but in two patients with cirrhosis, it was 2.9 g/dl (patient 10) and 1.9 g/dl (patient 4), or approximately 30% of plasma (Table 3).

In contrast, patients 12–15, with extrahepatic portal block (*i.e.*, isolated portal vein thrombosis), had no ascites despite marked portal hypertension and varix bleeding. In patient 15, ascitic fluid accumulated tran-



FIGS. 9A–D. Acute pylethrombosis in patient 33, with 2 weeks of abdominal pain. Despite resection of an "early" ischemic jejunal segment (*A*) and distal gastric resection of an unsuspected antral carcinoma (*C*), progressive small intestinal infarction led to death 25 days later. Closeup (*B*) of resected jejunum shows large venous clot, while abdominal computed tomography (after laparotomy) illustrates a mass of channels (*D*) with circumferential contrast (black arrowhead) consistent with varices from pylethrombosis.

TABLE 2. *Primary Clinical Manifestations of Pylethrombosis*

Etiology	Ascites*	Varix Hemorrhage	Bowel Infarction
Hepatic cirrhosis	++++	++++	+
Cavernous transformation (Extrahepatic portal block)	±	++++	±
Acute portomesenteric thrombosis (pylephlebitis)	±†	–	++++
Intraabdominal malignancy (without 1° liver disease)	++++	–	+

* Usually extremely low in protein content.

† Bloody exudate often accompanies hemorrhagic bowel, but the volume seldom is grossly detectable.

siently after a failed makeshift portasystemic shunt and its total protein was only 0.3 g/dl, less than five per cent of the plasma level.

Each of the other patients had varying amounts of intraperitoneal fluid, but it was rarely grossly detectable and the nature of the fluid conformed to the underlying pathophysiologic process. Thus, with hemorrhagic bowel infarction, there was uniformly intense edema and congestion of the digestive tract with free intraabdominal fluid invariably either deeply blood-tinged or frankly bloody and commonly foul-smelling from contamination by anerobic bacteria. When sampled at operation, this bloody fluid yielded total protein levels higher than 3.5 g/dl, or more than 50% of plasma. With septic pylephlebitis, intraabdominal fluid was also consistently high in protein content, commonly teeming with bacteria, and occasionally frankly purulent.

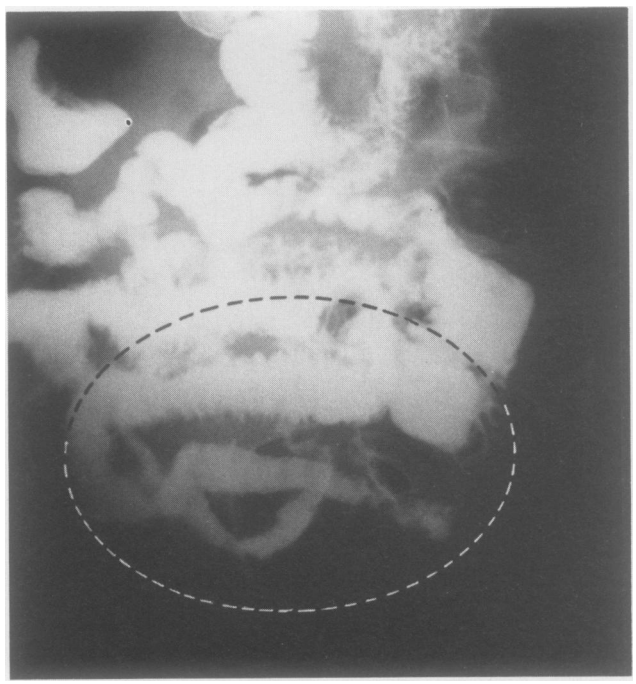


FIG. 10. Small bowel contrast roentgenogram in patient 22, with known portomesenteric venous thrombosis and persistent postprandial pain and anorexia. Note edema, spicules, "thumbprinting," and poor distensibility of distal ileum.

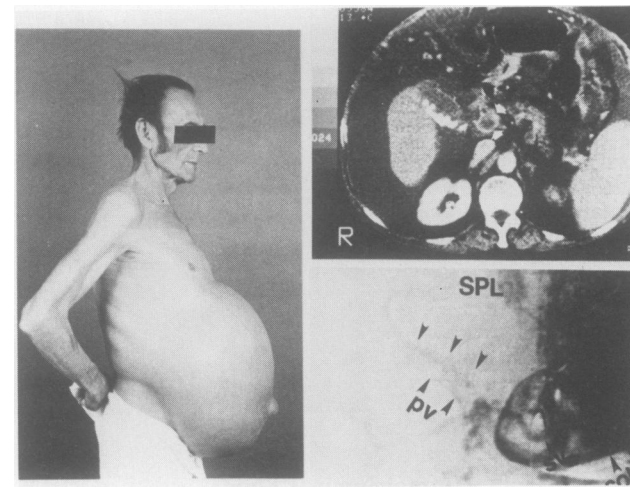


FIG. 11. Initial presentation of cirrhosis (Table 1, patient 10) with prominent ascites (left). One year after insertion of a LeVeen shunt, and 2 weeks after umbilical herniorrhaphy ascites recurred and became massive. Computed tomography (CT) with contrast enhancement (upper right) demonstrated only the outer wall of the portal vein consistent with portal vein thrombosis (white arrows). This finding was corroborated on digital-subtraction angiography (lower right), displaying filling defects in the portal vein and absence of venous arborization within the liver. SPL, splenic arterial injection; pv, portal vein; SV, splenic vein; and coll, venous collateral.

Discussion

Although pylethrombosis is potentially lethal, clinical presentations vary, depending on acuteness and extent of venous occlusion and associated complicating conditions. In cirrhosis, where longstanding restriction to transhepatic portal flow favors development of visceral phlebosclerosis and clot formation,[2] or where primary liver cell carcinoma encroaches on the portal vein,[3] pylethrombosis characteristically produces massive, intractable ascites, often accompanied by varix hemorrhage.[4] Propagation of thrombus into the mesenteric system occasionally leads to intestinal infarction,[1,5] but extensive venous collaterals minimize this complication or commonly limit it to a resectable segment of ileum or jejunum. Although mortality is high, varix hemorrhage with ascites is correctable by portal decompression (patient 1)[6]; intractable ascites without varix hemorrhage is correctable by insertion of a peritoneojugular shunt

(patient 10). Portal occlusion is highly suspect if ascites rapidly worsens or suddenly occurs with varix hemorrhage.[5] This is particularly true when there is no clear precipitating cause such as persistent alcoholism, dietary indiscretion, and noncompliance with diuretic drugs.

As in cirrhosis, patients with isolated portal block develop portal hypertension and varix rupture. Unlike cirrhosis, ascites is unusual despite equivalent portal hypertension,[7] probably because of: (1) normal intrahepatic portal pressure;[8] (2) more favorable edematogenic safety factors in the extrahepatic splanchnic microcirculation (*i.e.*, wider transcapillary oncotic gradient, high solute reflection coefficient and greater lymphatic capacity);[9] and (3) lack of renal salt and water retention,[10] factors that minimize the burden on visceral and diaphragmatic lymphatic drainage.[11] On the other hand, the magnitude of venous collateralization and the intensity of digestive tract congestion appear greater in cirrhosis than in isolated extrahepatic portal block despite equivalent portal hypertension (personal observations, CLW). This suggests other hemodynamic (*e.g.*, hyperkinetic splanchnic flow in cirrhosis[12]) or unknown factors. Indeed, in patient 22, with noncirrhotic pylethrombosis where venous clot had propagated into the superior mesenteric venous system, ascites was a persistent problem.

TABLE 3. *Ascitic Fluid and Plasma Protein Content in Pylethrombosis*

Patient	Etiology	Total Protein (g/dl) Ascitic fluid (AF)	Plasma (P)	Ratio AF/P (%)
2	Cirrhosis	0.1	6.2	2
3	Cirrhosis	0.3	4.9	6
4	Cirrhosis*	1.9	6.4	30
6	Cirrhosis	<0.1	6.5	2
7	Cirrhosis†	0.6	5.9	10
10	Cirrhosis	2.9	7.8	33
11	Cirrhosis	1.0	6.2	16
22	None	0.2	5.0	4
29	Ca pancreas	0.4	5.1	8
30	Ca pancreas	0.2	6.2	3
31	Ca pancreas	0.6	7.5	8
Mean		0.75	7.2	11
SD		0.88	0.94	11

* Also hepatoma.
† Also spontaneous bacterial peritonitis (*Escherichia coli*).

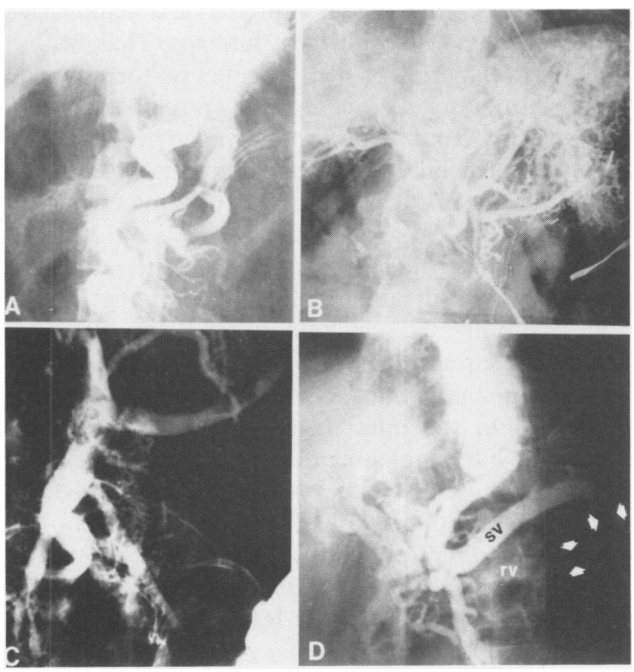


FIG. 12. Intraoperative portograms done *via* a jejunal (*A, C, D*) or omental (*B*) venous tributary. In *A* and *B*, chronic extrahepatic portal block was clinically suspected, but preoperative celiac-mesenteric arterioportography was inconclusive (patients 13 and 15). In *C* (patient 19), direct portography revealed not only complete occlusion of the portal vein but propagated thrombus in both the greater and lesser splanchnic circuits (see Fig. 6B). *D* confirms patency of a splenorenal anastomosis (arrows) immediately after construction.

The prognosis in patients with isolated portal vein block, in contrast to those with liver disease, is favorable even when portal decompression fails and hematemesis sporadically recurs.[13] Segmental portal decompression (*e.g.*, distal splenorenal shunt) may have theoretical advantages over standard portasystemic shunt (*e.g.*, proximal splenorenal or mesocaval) in extrahepatic portal block,[14] but the outlook is good after either operation.[15] Usually, the diagnosis is suspected by varix hemorrhage in the absence of liver disease, but with prior splenectomy and suboptimal visualization on splanchnic arterioportography direct intraoperative portography is confirmatory (Fig. 12). This technique not only corroborates portal vein occlusion but also establishes patency or occlusion of other portal trunks (mesenterosplenic) and a newly constructed portasystemic shunt.

In contrast to isolated portal vein occlusion with insidious portal hypertension and then varix rupture, acute portal thrombosis and suppurative endophlebitis generally lead to extensive clot propagation and widespread bowel infarction. Most cases are idiopathic,[16] but abnormal blood clotting is incriminated on occasion.[17-21] In three patients tested in this series (17, 18, and 22) blood antithrombin III levels were normal. Survival generally depends on prompt laparotomy and resection of infarcted bowel. In patient 22, however, persistent anorexia, chronic ascites, and distal small bowel ischemia led to ileal perforation and death 1 year later, while in patient 16, symptoms of abdominal pain subsided spon-

taneously. In this last individual, progressive portal hypertension and varix hemorrhage may yet develop, although 1 year after diagnosis of portal thrombosis he is well.

Differential diagnosis of acute pylethrombosis is particularly troublesome. Individuals may be young and often are mistakenly treated for viral gastroenteritis until persistent symptoms, findings of peritonitis, hematochezia, and hypotension supervene. Although plain abdominal roentgenograms are nonspecific, suspicious findings include small bowel dilatation with a paucity of colon air, thickened edematous bowel loops, and fluid sequestered in both bowel and peritoneal cavity. Whereas splanchnic arterioportography and computed tomography with contrast enhancement are diagnostic, life-threatening symptoms often require prompt operation. It is probably unwise to reconnect the bowel at the time of initial resection because the extent of venous occlusion and the viability of residual bowel are unclear. Indeed, intraoperative portography may uncover unsuspected clot propagation within gastrosplenic, mesenteroportal, and intrahepatic portal radicles (Fig. 12).

Suppurative portal endophlebitis (pylephlebitis) has the same symptoms as acute portal thrombosis although, like peripheral deep venous disease, the distinction between "phlebitis" and "thrombosis" is somewhat contrived. Nonetheless, the high death rate with pylephlebitis derives not only from ischemic bowel but also from ongoing intraabdominal sepsis. Thus, both perforated and nonperforated sigmoid diverticulitis, suppurative appendicitis, amebic colitis, and necrotizing pancreatitis, with or without metastatic intrahepatic abscesses, are life-threatening whether or not associated with portal thrombosis and bowel infarction. Whereas fulminant infection makes blood hypercoagulable,[22] "pyemic" injury to portal endothelium[23,24] initiates leukocyte-platelet aggregation with propagation of clot.[25] Although less easily identified (*e.g.*, viral infections), a similar pathogenesis may account for acute agnogenic portal thrombosis. In our experience, the value of early and prolonged anticoagulation[26] is difficult to assess as most patients either died quickly from uncontrolled sepsis or survived with or without anticoagulant therapy.

In the final group, portal occlusion accompanied an intraabdominal malignancy. Clinical manifestations typically were anorexia, weight loss, and progressive ascites, but one patient with a localized distal gastric carcinoma had severe abdominal pain from bowel ischemia (Trousseau's syndrome). The diagnosis of portal occlusion was usually determined at autopsy, but in the more recent two patients, portal occlusion was verified by computed tomography. Despite known blood hypercoagulability in patients with advanced cancer,[27,28] most of these terminally ill individuals demonstrated direct tumor invasion of the portal vein. Here, anatomic findings resembled isolated portal thrombosis, but ascites was prominent and hematemesis was absent. This difference probably relates to the underlying process and time sequences. Thus, in carcinomatous pylethrombosis, tumor also plugs lymph trunks particularly diaphragmatic leading to visceral lymphatic blockage.[29] Overproduction of splanchnic filtrate from portal hypertension and greater microvascular permeability from tumor angiogenesis,[30,31] on the one hand, and impaired tissue fluid absorption from lymphatic blockade, on the other, together favor formation of ascites. This pathogenetic explanation is particularly well-illustrated by the clinical course of patient 34, who developed rapidly progressive and intractable ascites 4 months after diagnostic laparotomy for pancreatic carcinoma with diffuse intraabdominal metastases. At autopsy 6 weeks later, in addition to widespread lymphangiocarcinomatosis, the portal vein was found to be occluded by tumor thrombus. Early death, on the other hand, precludes development and rupture of esophageal varices, which takes many months or even years after simple portal vein occlusion.[32]

Although not systematically chronicled in this review, sporadic experience with acute portal occlusion that occasionally follows splenectomy for hematologic disorders and distal splenorenal shunt for varix hemorrhage in cirrhosis conforms with these findings. Thus, with a normotensive portal system, acute portomesenteric thrombosis after splenectomy typically precipitates diffuse ileojejunal infarction. Comparable venous occlusion of the greater splanchnic circulation after distal splenorenal shunt is characteristically heralded by progressive, refractory ascites.

In view of this paradox, it is worthwhile to reexamine the pathogenesis of ascites in pylethrombosis. In the acute syndromes, bloody fluid characteristically accumulates as plasma and red blood cells extravasate in bulk from intensely congested and disrupted intestinal capillaries. On the other hand, intraabdominal fluid associated with carcinomatous or "cirrhotic" portal thrombosis is usually watery or light straw-colored. Of the 11 patients studied, nine exhibited extremely low protein ascitic fluid (protein washdown). This finding is consistent with increased movement of salt and water out of proportion to protein leakage in response to extrahepatic portal microvascular hypertension.[33,34] The protein concentration of mesenteric tissue fluid and lymph progressively declines, and ascitic fluid low in protein "transudes" from the serosa of the bowel and mesentery. This unusual peritoneal transudate has been observed primarily in portal occlusion complicating cirrhosis, locally-invading carcinoma, thrombosed or inef-

fective portacaval shunts, and far-advanced cirrhosis where the portal vein, though anatomically patent, is nonfunctional.[35,36] In two cirrhotic patients, ascitic fluid protein was approximately 30% of plasma, perhaps reflecting incomplete mixing with excess hepatic lymph. The appearance of extremely low protein intraabdominal fluid relative to plasma, therefore, should raise the suspicion of portal occlusion, not only in patients with cirrhosis and intraabdominal carcinoma, but also in isolated extrahepatic portal block (where ascites is rare) and following portasystemic shunt where the anastomosis is occluded or ineffective, and portal hypertension persists.

The diagnosis of pylethrombosis rests not only on recognition of its protean clinical manifestations but also on accurate imaging. Splenoportography is the classical radiographic method and, although still useful, has been supplanted by safer and more revealing splanchnic arterioportography and its newer digital-subtraction-computed modification. On occasion, intraoperative portography with its vivid anatomic definition is indispensable. Other imaging techniques include real-time ultrasonography and computed tomography. Whereas ultrasound has been recommended to screen for pylethrombosis,[37-39] portal images are often indefinite especially with massive ascites, obesity, and meteorism.[40] Moreover, they cannot be enhanced by contrast infusion. On the other hand, computed tomography—especially with contrast enhancement—not only accurately delineates pylethrombosis[41] but also provides information about possible coexistent periportal tumor,[42] abscess, or other intraabdominal perturbations. Arterioportography and computed tomography are now rightfully considered the mainstays for documenting portomesenteric venous occlusion and, in general, are complementary both for diagnosis and for defining therapeutic options. Despite their success, however, other noninvasive and perhaps more accurate methods are already on the horizon. For example, magnetic resonance imaging, which entails no irradiation, provides an intensely dark image of the portal system even sharper than in computed tomography.[43] As yet, however, it is neither widely available nor extensively tested.

In summary, clinical symptoms and signs of pylethrombosis depend on the acuteness and extent of visceral venous blockade, the severity of portal hypertension, auxiliary venous collateralization, and visceral lymph flow. The primary inciting factors are: endothelial damage and blood hypercoagulability induced by trauma, infection, stagnant circulation, blood dyscrasia, and malignancy. Improved imaging techniques now allow more accurate and earlier recognition and offer the prospect of earlier effective treatment of this catastrophic disorder.

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