Pneumatosis Intestinalis

Surgical Management and Clinical Outcome

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Pneumatosis intestinalis (PI) occurs in a wide variety of patients, some of whom require urgent surgery, while others can be observed with resolution of symptoms and radiographic findings. During 1 year, 27 patients with PI were prospectively evaluated for clinical, laboratory, and radiographic features that would be useful in predicting the need for surgery, the pathologic findings, and patient outcome. Sixteen of the twenty-seven patients underwent laparotomy, with only one negative exploration. Of the 11 patients not explored, there were two deaths in moribund patients. Seven of nine patients with jejunostomy tubes, recent gastrointestinal anastomoses, inflammatory bowel disease, lactulose therapy, or chemotherapy who did not have clinical evidence of an acute surgical abdomen or metabolic acidosis survived without surgery (two deaths unrelated to the gastrointestinal tract). Patients presenting with bowel obstruction and PI required surgery in seven of nine cases, did not have necrotic bowel, and had 11% mortality. Eight patients with ischemic bowel had a 75% mortality rate, despite surgery. Patients with PI and clinical evidence of bowel obstruction or ischemia usually require urgent surgery, while asymptomatic patients without metabolic acidosis can be safely observed.

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PNEUMATOSIS INTESTINALIS (PI) is defined as the presence of gas in the bowel wall and can be classified as either primary of secondary. Primary PI radiologically and pathologically consists of cystic collections of air, usually located in the colon. It is generally a benign condition that may remain stable for years but may be complicated by obstruction or bleeding. Its etiology is unknown, but its frequent association with emphysema suggests that air may dissect through the mediastinum and retroperitoneal tissues to enter the bowel mesentery and subserosal spaces.[1] Secondary PI, on the other hand, is characterized by linear or circumferential air in any part of the gastrointestinal tract and is associated with many clinical conditions. A review of 213 cases in 1952 by Koss included a 15% incidence of primary PI compared to 85% of cases that were secondary to other underlying disorders.[2] Pneumatosis intestinalis may be associated with pneumoperitoneum either as a benign condition or following perforation of a viscus. In addition PI may be associated with portal venous gas, which is often an ominous sign and has a reported mortality rate of 75%.[3,4]

The first pathologic description of PI is attributed to DuVernoi in 1730 during a cadaver dissection.[5] In the 18th century John Hunter contributed two specimens of hog intestine to the Museum of the Royal College of Surgeons that demonstrated multiple gas-filled cysts beneath the serosal layer.[6] Bang[7] in 1876 published the first comprehensive description of PI in a human, including microscopic features. Finney[8] published the first description of PI in the United States in 1908. This case was diagnosed at surgery in a patient with an obstructing gastric carcinoma. Finney believed that PI itself represented a distinct and very rare tumor, a theory now discredited. Gazin et al.[9,10] reported the first case diagnosed before operation by x-ray and confirmed by surgery and histologic study.

Although more than 350 case reports of PI have been published, there is a lack of information regarding the clinical management and predictors of outcome of these patients. Managing patients with PI can be challenging because urgent surgery is required in those patients with bowel infarction. On the other hand, many patients with PI have a benign course and can safely avoid surgery. A prospective evaluation of all patients with PI at Duke University Medical Center and the Durham VA Medical Center was therefore performed during a 1-year period to obtain information that would prove to be useful in management decisions.

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Methods

During a period of 1 year at Duke University Medical Center and the Durham VA Medical Center, there were 27 patients with radiographic evidence of PI. One patient had two episodes of PI with radiographic resolution between them. Another patient had three episodes with the first two treated with bowel resection. However, for the purpose of this study, each patient was counted only once. Newborns with necrotizing enterocolitis were excluded from study because this appears to be a distinct clinical entity. All x-rays were reviewed by a single gastrointestinal radiologist (RPR). To examine the correlation between clinical findings and the need for surgery and clinical outcome, six parameters of the history and physical exam were graded as either absent, mild, moderate, or severe (0, 1, 2, or 3+). These included pain, diarrhea, fever, tenderness, blood per rectum, and hypotension. The sum of these scores served as an index of the severity of the patient's clinical condition. In addition laboratory values were obtained on the day of diagnosis of pneumatosis, including WBC, pH, bicarbonate, AST, ALT, alkaline phosphatase, lactic acid, and amylase.

Patients were entered into the series at the time of the initial radiographic diagnosis of PI. At the end of 1 year, the films were all reviewed and evaluated in terms of the documented duration of the PI, the distribution of the PI, whether the PI was linear and tangential to the bowel wall or circumferential, the degree of PI (1, 2, or 3+), whether diagnosed on plain films of the abdomen or CT or both, and whether there was portal venous gas in addition to gas in the bowel wall.

Clinical and radiographic features of the 27 patients were analyzed for correlation with the presence or absence of necrotic bowel and outcome.

Results

There were 13 male and 14 female patients. The mean age of the group was 51 years, with an age range of 1 month to 83 years. Most patients were in their fifth to eighth decade of life (Fig. 1).

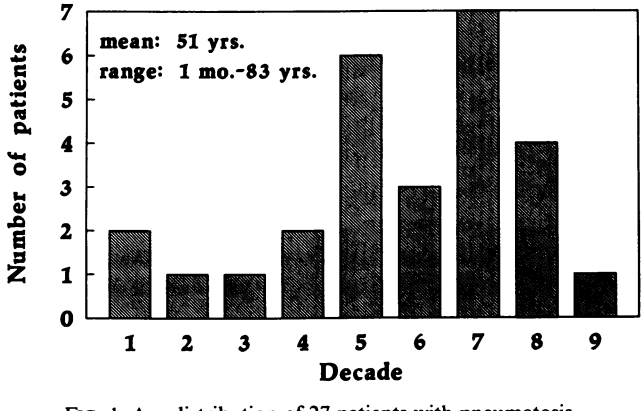

FIG. 1. Age distribution of 27 patients with pneumatosis.

TABLE 1. *Diagnoses of 27 Patients with Pneumatosis Intestinalis*

1. Bowel obstruction	9
Gastrointestinal tumors	5
Chronic peptic ulcer	1
Fecal impaction	1
Ileus without mechanical obstruction	2
2. Ischemic bowel	8
Hypotension	4
Mesenteric thrombosis	2
Mesenteric emboli	1
Mesenteric trauma	1
3. Trauma to bowel wall	3
4. Hepatitis/hepatic cancer	3
5. Cancer chemotherapy	2
6. Inflammatory bowel disease	1
7. Primary pneumatosis intestinalis	1

The principal diagnoses of the patients are listed in Table 1. Nine patients presented with bowel obstruction. Five of these patients had known gastrointestinal tumors (3 colonic, 1 gastric, and 1 pancreatic). All five underwent surgery for relief of obstruction, and none died perioperatively. One patient with known peptic ulcer disease and Alzheimer's disease presented with gastric outlet obstruction, gross gastric pneumatosis, and gross intrahepatic portal venous air by computed tomography. After a course of nasogastric suctioning and antibiotics, his PI and gastric outlet obstruction resolved. Two patients presented with colonic dilation and PI, one due to severe fecal impaction and the other due to prolonged colonic ileus. Both were explored, found to have viable bowel, and underwent decompressive colostomy.

Eight patients had a diagnosis of ischemic bowel confirmed by surgery. The causes of their intestinal ischemia are listed in Table 1. Only two of these patients survived. Three trauma patients developed PI radiographically after laparotomy and repair of bowel injuries. All three survived without additional surgery. The only death in a trauma patient occurred in a child who presented with severe hypotension and acidosis and at laparotomy was found to have a mesenteric tear resulting in bowel necrosis.

Sixteen of the twenty-seven patients underwent exploratory laparotomy. Nine of these procedures involved bowel resection, usually for ischemic bowel (Table 2). An

TABLE 2. *Surgical Procedures Performed*

1. Bowel resection	
for ischemic bowel	7
for colitis	1
for recurrent tumor	1
2. Bypass of obstructing tumor	3
3. Colostomy	2
4. Open and close (not salvageable)	1
5. Negative exploratory laparotomy	1
Total	16

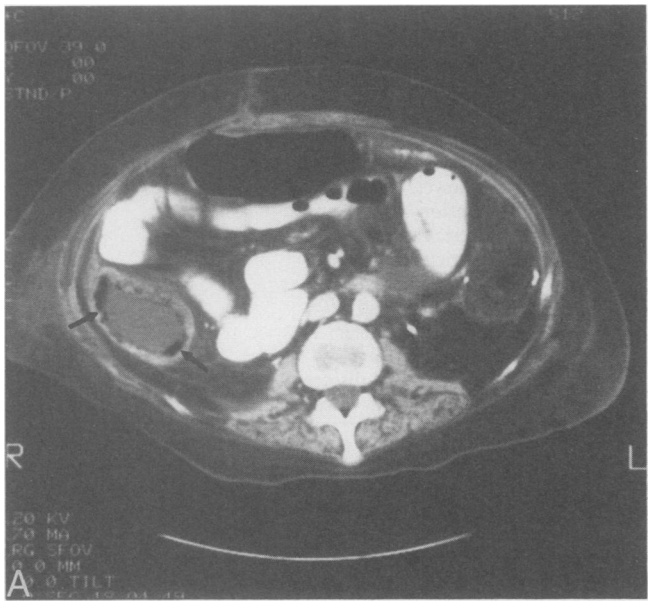

FIG. 2A. Computed tomography demonstrates pneumatosis of the right colon (arrows).

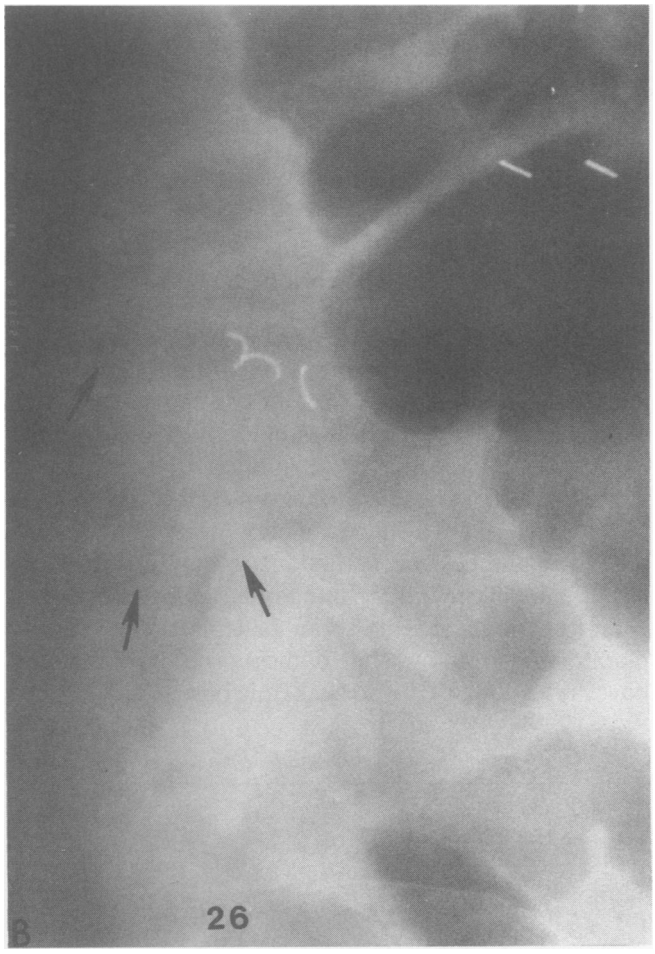

FIG. 2B. Plain film the following day shows subtle (1+) pneumatosis of right colon (arrows). At surgery the same day there was no demonstrable bowel pathology. The pneumatosis resolved after operation.

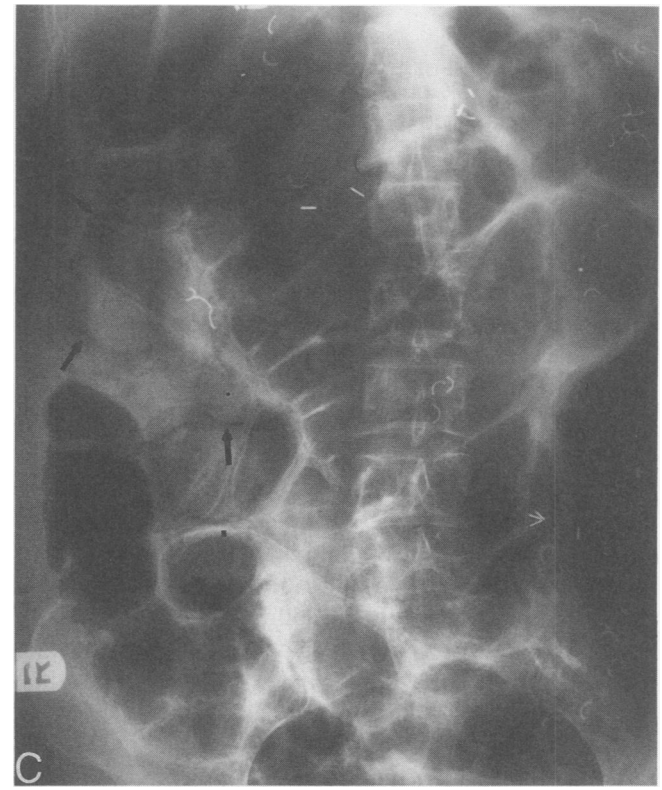

FIG. 2C. Plain film 2 weeks later again shows pneumatosis (3+) associated with gross ileus but no other significant complaints. The patient was discharged without further surgery.

obstructing tumor was bypassed in three patients. One patient was found to have necrotic bowel from the ligament of Treitz to the anus and was therefore closed and died within 24 hours. Only one patient had no pathologic findings at laparotomy despite obvious PI by plain films and CT scan (Fig. 2A and B). This patient had undergone enterolysis for small bowel obstruction 5 days earlier and also carried a diagnosis of *melanosis coli*. Her PI resolved following re-exploration but recurred 2 weeks later (Fig. 2C). The second episode was treated without operation with radiographic and clinical resolution.

Nine of the twenty-seven patients in this series died, for an overall mortality of 33% (Table 3). Eleven patients observed without surgery had a mortality rate of 18%, while those undergoing surgery had a mortality rate of 44%. The highest mortality rate was seen in patients who at surgery were found to have necrotic bowel. Six of these eight patients died perioperatively. In contrast all six patients with gastrointestinal tumors and PI required surgery and none died perioperatively. Among five patients with feeding jejunostomy tubes, there was one death due to ischemic bowel. Although a jejunostomy tube is often associated with benign PI, the patient who died was also septic, hypotensive, and died due to bowel infarction. Of the three postoperative cardiac surgical patients explored

TABLE 3. *Mortality by Associated Condition**

Associated Condition	N	Died	% Mortality
GI tumor	6	0	0
Bowel obstruction	9	1	11
Jejunostomy tube	5	1	20
Trauma	4	1	25
Chemotherapy	2	1	50
Status after cardiac surgery	3	2	66
Ischemic bowel	8	6	75
Surgery	16	7	44
No surgery	11	2	18
Overall	27	9	33

* Some patients may be included in more than one category.
GI, gastrointestinal.

for PI, one died of necrotic bowel related to prolonged hypotension, and another died due to multisystem failure not directly related to her gastrointestinal problems. One of two patients with PI who was being treated with chemotherapy died, but death was due to brain metastasis. Two patients with hepatitis who were receiving lactulose developed PI. Neither developed clinical signs of an acute abdomen, and neither required surgery.

Of the two patients who died without surgery, one had known lung cancer metastatic to brain and his family requested that no surgery be performed. Autopsy was not done in this case and the viability of the bowel was therefore not determined, although necrosis was strongly suspected. The second nonsurgical death occurred in a patient with fulminant hepatitis who died of liver failure. Autopsy showed normal bowel. The nine patients who improved without surgery did not manifest any clinical signs that would have prompted laparotomy.

Indices of severity of illness correlated closely with clinical outcome. No patient with a severity of illness score of less than 4 had necrotic bowel and all survived. Patients with an intermediate score[5-7] had necrotic bowel in two of seven cases, and six of seven survived. Patients who appeared by history and physical exam to have acute abdominal crises, with a total score 8 or more had the highest incidence of necrotic bowel, and only three of nine survived (Table 4).

Acidosis with a pH of less than 7.3 was associated with necrotic bowel in five of six patients, three of whom died. Only 2 of 14 patients with a pH of more than 7.4 had necrotic bowel. Similarly metabolic acidosis indicated by serum bicarbonate level less than 20 was predictive of necrotic bowel in 7 of 10 cases with a 40% survival. This is in contrast to patients with bicarbonate level of more than 20, who fared better, with only 1 of 16 cases having necrotic bowel (Table 4). An elevated lactic acid level also correlated with necrotic bowel and diminished survival compared to patients with a normal serum lactate level. Finally hyperamylasemia of more than 200 occurred in three patients and all three had necrotic bowel. However most patients with necrotic bowel did not develop hyperamylasemia. The WBC, AST, ALT, and alkaline phosphatase levels had virtually no predictive value for the presence of dead bowel or for survival.

Radiographic Features of Pneumatosis

The duration of radiographically demonstrable PI varied from 1 to 21 days. Thirteen patients had PI that was demonstrable on only one day, although in most cases there were multiple films documenting the radiologic diagnosis. Three patients had documented PI for 2 days and another three patients had it for 3 days. Seven patients had demonstrable PI for 6 days or longer and each of these patients survived without surgical resection of the involved bowel.

In general the distribution of PI corresponded with the abnormality in those patients who had surgery. Fifteen patients had gross (3 plus) PI (Fig. 3). In only two of these patients was ischemia the proved etiology for the PI. In another three patients ischemia may have played a role. One of these patients had a decompressive cecostomy for a grossly distended cecum; another patient died without surgery but had severe hypotension associated with status epilepticus and generalized metastatic disease; a third patient was explored without demonstrable bowel infarction, but subsequent barium examinations suggested the probability of intestinal ischemia without full-thickness infarction. The remaining 10 patients with gross PI survived the episode without surgical resection.

In four patients the PI was rather subtle (1 plus). Three of these patients had surgical confirmation of infarcted bowel with resection, and the fourth patient had confirmation of the presence of PI by computed tomography.

TABLE 4. *Correlation of Clinical and Laboratory Values with Presence of Ischemic Bowel and Outcome*

	Necrotic Bowel	Survival
Clinical Severity Score		
<4	0/10	10/10
5–7	2/7	6/7
≥8	6/9	3/9
pH		
>7.4	2/14	10/14
7.3–7.4	1/2	1/2
<7.3	5/6	3/6
HCO_3 (mmol/L)		
<20	7/10	4/10
≥20	1/16	14/16
Lactic Acid (mmol/L)		
≤2	1/8	6/8
>2	4/7	3/7
Amylase (Iu/L)		
<200	4/21	18/21
>200	3/3	1/3

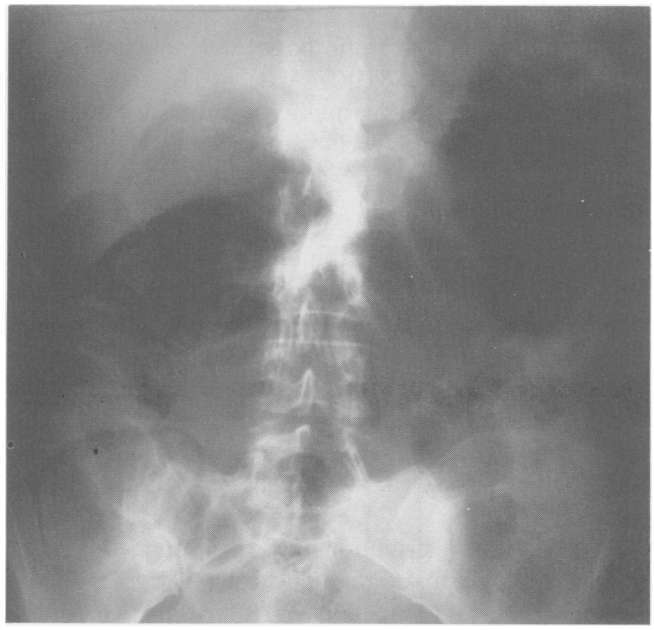

FIG. 3. Abdominal radiograph demonstrates gross linear pneumatosis (3+) involving most of the colon. This persisted for 2 weeks without significant abdominal symptoms. The patient was on lactulose for toxic hepatitis secondary to chemotherapy for malignant melanoma.

In this patient PI persisted for 8 days and was associated with chemotherapy and partial bowel obstruction.

Pneumatosis intestinalis is most conspicuous when a linear collection of gas runs tangentially along the bowel wall. Circumferential PI following the course of the valvulae conniventes may be less obvious radiographically (Fig. 4). Fourteen patients had a linear distribution of gas only, four patients had circumferential lucencies only, and in eight patients there were radiographic findings of both linear and circumferential PI. In one patient the PI was demonstrable by CT only. This patient had a gastrostomy tube and multiple enterotomies with postoperative ileus following a gunshot wound. A plain film of the abdomen 1 day after the computed tomographic examination did not demonstrate PI. Computed tomography is more discriminating in the recognition of subtle gas lucencies than are plain radiographs.[11]

Portal venous gas may or may not be present in association with PI. In this series of 27 patients with PI, there were 11 patients in whom there was demonstrable portal venous gas. Four patients had definite ischemic disease with infarcted bowel resected surgically, two patients probably had ischemic disease, and the remaining five patients had definite portal venous gas but survived the episode without surgical resection. Two of these patients had gross gastric pneumatosis associated with gastric outlet obstruction and gross portal venous gas. Both the gastric pneumatosis and the portal venous gas resolved promptly with nasogastric tube decompression of the stomach.

Discussion

Two basic mechanical features characterize the majority of cases of PI: mucosal injury, the most important and prevalent feature, and increased intralumenal pressure. Increased intralumenal pressure may be produced by bowel obstruction, ileus, or iatrogenically by upper or lower GI endoscopy.[12,13] Potential causes of mucosal injury are multiple and most reports of PI involve one or more of these agents. The mucosa of the GI tract is highly sensitive to ischemia and mesenteric ischemia of any etiology may result in PI.[14] Enteritis of viral, bacterial, or fungal etiology may precede its onset.[15,16] Gas-forming organisms invading the bowel wall may produce PI. Iatrogenic trauma to the gastrointestinal mucosa from either sclerotherapy, endoscopy, placement of a feeding jejunostomy tube, or surgical trauma from a gastrointestinal anastomosis may be associated with PI.[13,17,18] Inflammation of the mucosa secondary to inflammatory bowel disease, graft-*versus*-host disease, and connective tissue

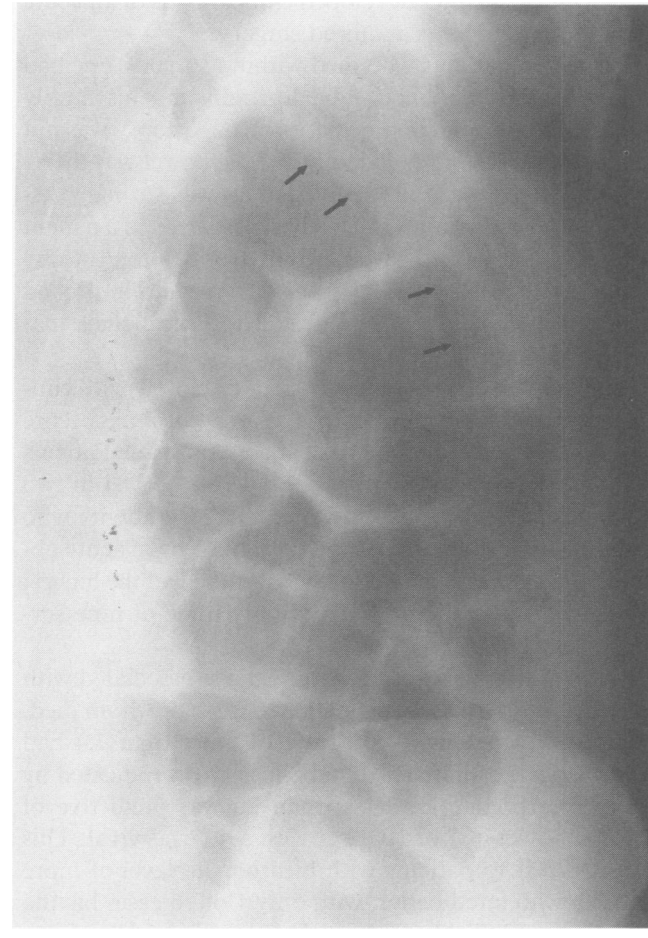

FIG. 4. A coned radiograph of the left upper quadrant demonstrates subtle (1+) circumferential pneumatosis of jejunal segment. At surgery most of the jejunum and left colon were infarcted. The patient died 7 days after resection.

diseases may be associated.[13,19,20] Radiation enteritis or ingested corrosive agents may damage mucosa.[13] Multiagent chemotherapy, high-dose steroid administration, lactulose, and bone marrow transplantation have all been implicated.[21-23] Ulceration, either of the benign peptic form or by malignant gastrointestinal tumors, may precede PI. Sometimes PI may be due to pulmonary alveolar rupture with dissection of gas along retroperitoneal routes to the bowel wall. Only 1 of the 27 cases in our series lends itself to this explanation.

The frequent occurrence of PI with 27 cases at one medical center within 1 year suggests that the condition is more common than generally appreciated and that it may be underdiagnosed. Surgical consultation is frequently sought for the patient with either symptomatic or asymptomatic PI. Because the condition ranges from a benign condition requiring no specific therapy to a grave clinical problem requiring emergent surgery, factors that correlate with outcome may be useful to the clinician.

In this series PI was evenly distributed throughout the gastrointestinal tract and location did not correlate with gravity of disease. Presence of gross PI usually implied a benign clinical course. The presence of portal venous gas was associated with 37% mortality.

Patients with a radiographic diagnosis of PI should have a careful history and physical examination, which, along with knowledge of the patient's underlying disease, provide most of the information regarding need for exploratory laparotomy. Generally patients with severe abdominal pain or tenderness associated with diarrhea, blood per rectum, fever, or hypotension had abdominal surgery in this series. In addition the presence of metabolic acidosis, particularly lactic acidosis, and markedly elevated serum amylase suggest the presence of ischemic bowel. Patients whose preoperative diagnosis suggested ischemia as the primary cause of PI were more likely to have bowel necrosis than those whose PI resulted from bowel obstruction. Patients with ischemic bowel had the poorest prognosis, but the 25% salvage rate supports the role for aggressive surgery in these patients. Patients with gastrointestinal tumors or obstruction did well with surgery.

Pneumatosis intestinalis may occur after mucosal injury of any etiology, often combined with increased intraluminal pressure. Patients manifesting symptoms or signs of abdominal distress merit an aggressive surgical approach. Associated acidosis and hyperamylasemia suggest necrotic bowel and portend a grave prognosis. The finding of PI radiographically without associated clinical evidence of intra-abdominal pathology does not necessitate operative exploration. These patients may be followed expectantly.

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