Pneumatosis intestinalis

AUTHORS: Eric Goldberg, MD, J Thomas Lamont, MD
SECTION EDITOR: Lawrence S Friedman, MD
DEPUTY EDITOR: Shilpa Grover, MD, MPH, AGAF

All topics are updated as new evidence becomes available and our peer review process is complete.

Literature review current through: **Sep 2023.**
This topic last updated: **Feb 01, 2023.**

INTRODUCTION

Pneumatosis intestinalis (PI) refers to the presence of gas within the wall of the small or large intestine. Intramural gas can also affect the stomach, but this condition is referred to as gastric pneumatosis [1]. Since its first description, PI has appeared in the literature under many names, including pneumatosis cystoides intestinalis, intramural gas, pneumatosis coli, pseudolipomatosis, intestinal emphysema, bullous emphysema of the intestine, and lymphopneumatosis [2,3]. PI can also be associated with pneumoperitoneum or free air in the peritoneal cavity.

The pathogenesis of PI is poorly understood and is probably multifactorial. In some cases, PI is an incidental finding associated with a benign etiology, whereas in others, it portends a life-threatening intra-abdominal condition. As a result of the wide array of clinical settings in which PI is encountered, its implications are often misinterpreted. This topic will review the epidemiology, pathogenesis, clinical features, evaluation, and management of PI. The clinical features of necrotizing enterocolitis, an important cause of PI in newborns, is discussed in detail separately. (See "Neonatal necrotizing enterocolitis: Clinical features and diagnosis".)

EPIDEMIOLOGY

The incidence of pneumatosis intestinalis (PI) is difficult to ascertain since most patients are asymptomatic and never come to clinical attention [3]. Adults are typically diagnosed in the fifth

to eighth decade. (See "Neonatal necrotizing enterocolitis: Clinical features and diagnosis".)

ETIOLOGY AND PATHOGENESIS

Pneumatosis intestinalis (PI) is idiopathic (15 percent) or secondary (85 percent) to a wide variety of gastrointestinal and non-gastrointestinal illnesses (table 1) [4,5]. The majority of cases in infants are secondary to necrotizing enterocolitis. (See "Neonatal necrotizing enterocolitis: Clinical features and diagnosis".)

Numerous hypotheses have been proposed to explain the pathogenesis of PI, including mechanical, bacterial, and biochemical causes. Although the theories are distinctly different, they are not necessarily mutually exclusive. It is likely that multiple pathogenic mechanisms are involved in the formation of PI.

- **Mechanical theory** – According to the mechanical theory, gas dissects into the wall of the bowel from either the luminal surface through breaks in the mucosa or through the serosal surface by tracking along mesenteric blood vessels [6]. Once inside the bowel wall, gas may spread along the mesentery to distant sites [7].

  The mechanical theory has gained support for several reasons. First, PI can be reproduced experimentally by insufflating air into an excised colonic segment in which mucosal incisions have been made [8]. Second, the mechanical theory accounts for the association of PI with conditions that disrupt mucosal integrity, such as necrotizing enterocolitis, intestinal ischemia, caustic ingestions, inflammatory bowel disease, and intestinal infections [7]. Third, the mechanical theory offers an explanation for the association of PI with obstructive pulmonary disease. In these patients, coughing may cause alveolar rupture and air may subsequently track along blood vessels into the mediastinum, through the diaphragm, and ultimately to the mesenteric root [9]. Once air has gained access to the mesenteric root, it may course along small mesenteric blood vessels that ultimately penetrate through the bowel wall.

  The argument against the mechanical theory is that the direct tracking of air from the lungs into the bowel wall is the invariable absence of a "gas trail" from the mediastinum to the intestine [10]. Furthermore, some studies have found that the gas composition of the intramural cysts does not resemble alveolar air, but rather, the cysts contain a different mixture of hydrogen, nitrogen, and carbon dioxide [11,12].

- **Bacterial theory** – According to the bacterial theory, PI results from gas-forming bacteria gaining access to the submucosa through breaches in the mucosa. In support of this

theory, PI has been reproduced experimentally by injecting the gas-forming bacillus *Clostridium perfringens* into the bowel wall of rats [13]. Further supporting the bacterial theory is that PI may resolve following treatment with antibiotics [14]. Finally, elemental diets have been reported to improve PI presumably by removing substrate for the production of gas by bacteria [15]. The argument against the bacterial theory is that the cysts are sterile and if they rupture, the resulting pneumoperitoneum often follows a benign course without development of peritonitis [16,17]. (See 'Antibiotics' below.)

- **Biochemical theory** – The biochemical theory proposes that luminal bacteria produce excessive amounts of hydrogen gas through fermentation of carbohydrates and other food. As the pressure of the gas within the intestinal lumen increases, gas may be forced directly through the mucosa and become trapped within the submucosa [18]. Indeed, the hydrogen content of cysts has been reported to be as high as 50 percent, and some patients with PI have much higher levels of breath hydrogen than controls [11,18,19]. PI has been reported in patients with small bowel bacterial overgrowth and in patients taking alpha glucosidase inhibitors and lactulose, which increase intestinal gas [20-24]. (See "Small intestinal bacterial overgrowth: Etiology and pathogenesis", section on 'Pathophysiology' and "Alpha-glucosidase inhibitors for treatment of diabetes mellitus".)

PATHOLOGY

Pneumatosis intestinalis (PI) can affect any portion of the gastrointestinal tract distal to the stomach. In one series that included 919 patients with PI, the small bowel, colon, or both were involved in 42, 36, and 22 percent of cases, respectively [25]. The cysts may be confined to the mucosa, submucosa, or subserosa, or involve all three layers. Subserosal cysts are more commonly seen in small intestinal pneumatosis, while submucosal cysts are more commonly seen in colonic pneumatosis [25].

On gross appearance, submucosal cysts are polypoid with the overlying mucosa displaying a bluish hue. Subserosal cysts are characteristically found near the mesenteric border adjacent to blood vessels and appear as multiple, glistening, pale-bluish, gas-filled blebs [16,26]. The cysts of PI are pseudocysts since they lack an epithelial lining. However, they may become surrounded by a rim of histiocytes, multinuclear giant cells, lymphocytes, neutrophils, eosinophils, granulomas, and fibrosis, especially after they collapse [18,27].

CLINICAL FEATURES

Clinical presentation — Most patients with pneumatosis intestinalis (PI) are asymptomatic and probably never come to clinical attention [3]. Patients who come to clinical attention present with symptoms related to either the presence of PI such as abdominal pain, obstruction, bleeding, or due to the underlying disorder associated with PI.

Symptoms associated with PI depend upon the region of intestine affected (table 2). In one review of 919 patients, the most common symptoms in patients with small intestinal pneumatosis were vomiting, abdominal distention, weight loss, abdominal pain, and diarrhea [25]. The most common symptoms in patients with colonic pneumatosis were diarrhea, hematochezia, abdominal pain, abdominal distention, and constipation. Other symptoms reported with PI include flatulence, loss of appetite, and tenesmus.

Complications — Complications of PI occur in approximately 3 percent of patients [26]. Complications include small and large bowel obstruction due to cyst encroachment on the lumen, volvulus, intussusception, or adhesions following cysts collapse and hematochezia due to ulceration of the mucosa overlying the cysts. Pneumoperitoneum may occur due to rupture of subserosal cysts.

Imaging findings — Findings suggestive of PI may be present on several imaging modalities.

- **Abdominal radiographs** – PI is often detected by plain films of the abdomen, which will be positive in approximately two-thirds of affected patients (image 1) [16,25]. Intramural gas can be linear, curvilinear, or circular in appearance [28,29]. Pneumoperitoneum may be detected on abdominal radiographs in 9 percent of patients [25].

  There may be a learning curve in the ability of radiologists to detect PI. In a study of 463 radiology trainees shown plain films with pneumatosis, only 28 percent could correctly identify PI. Increased years of training correlated positively with the ability to identify PI [30].

- **Abdominal ultrasound** – PI may be visible as high-amplitude gas echoes with acoustic shadowing [31].

Endoscopic findings — PI may be discovered incidentally on screening sigmoidoscopy or colonoscopy and prompt further imaging with endoscopic ultrasound [32]. Cysts vary in size from a few millimeters to several centimeters. Submucosal cysts usually have a pale or bluish appearance and when biopsied, can rapidly deflate with an audible hiss [10,33]. Biopsy of the submucosal cysts can be diagnostic as the blebs can be confused with other lesions. The authors typically biopsy the submucosal cysts as the deflation following biopsy is diagnostic and

there is minimal clinical risk in the maneuver. If PI is suspected, cross sectional imaging is routinely performed to see the distribution and extent, assess for underlying associations, and rule out other ominous features such as portal venous gas.

EVALUATION

Pneumatosis intestinalis (PI) is usually detected on imaging or endoscopy performed for evaluation of abdominal symptoms. Assessment of patients with suspected PI should include a history, physical examination, laboratory studies, and abdominal imaging. We perform a contrast-enhanced abdominal computed tomography (CT) scan to establish the diagnosis, determine the underlying etiology, and diagnose associated complications. Magnetic resonance imaging can also be used, but it is more time consuming, and is less widely available [34].

History and physical examination — The patient's history should be reviewed to identify the underlying etiology of PI with particular attention to risk factors for intestinal ischemia and obstruction (table 1). In the absence of complications, physical examination of the abdomen is normal but in some cases the abdomen may be mildly distended. A mass can be palpated on abdominal or digital rectal examination in approximately 10 percent of patients [25,35]. In rare cases, PI can simulate organomegaly [36]. In patients with intestinal ischemia or bowel obstruction, the abdomen is grossly distended and peritoneal signs may be present. (See "Overview of intestinal ischemia in adults", section on 'Risk factors' and "Etiologies, clinical manifestations, and diagnosis of mechanical small bowel obstruction in adults", section on 'Etiologies' and "Large bowel obstruction", section on 'Etiology'.)

Abdominal CT scan — Characteristic findings of PI on abdominal CT scan include circumferential (cystic) collections of air adjacent to the lumen of the bowel that run in parallel with the wall of the bowel or linear collections without the air-contrast or air-fluid levels characteristically seen with intraluminal air (image 2) [37,38]. Decreased mural contrast-enhancement and the presence of associated portal venous gas in patients with PI are suggestive of intestinal ischemia [39]. However, 30 percent of patients with PI and portal venous gas have a benign idiopathic cause [40].

Findings on abdominal CT that are not predictive of clinically worrisome PI include the presence of free peritoneal air (pneumoperitoneum) and linear versus cystic morphology [41]. (See "Mesenteric venous thrombosis in adults", section on 'Diagnosis' and "Nonocclusive mesenteric ischemia", section on 'Plain abdominal films' and "Overview of intestinal ischemia in adults", section on 'Plain radiographs'.)

In patients with PI and clinical suspicion of mesenteric ischemia, presence of the pneumatosis in the colon may portend a worse prognosis. In a retrospective study of 540 patients with suspected mesenteric ischemia referred for CT, multivariate analysis showed an increased short-term mortality in patients with colonic PI [42].

Laboratory studies — Laboratory studies are usually normal in patients with PI. Findings suggestive of the presence of mesenteric ischemia or bowel infarction include a marked leukocytosis with a predominance of immature white blood cells, an elevated hematocrit consistent with hemoconcentration, and metabolic acidosis with serum lactate >2.0 mmol/L. (See "Overview of intestinal ischemia in adults", section on 'Laboratory studies'.)

DIFFERENTIAL DIAGNOSIS

Stool can mimic the appearance of pneumatosis intestinalis (PI) on plain abdominal films. Mucosal (eg, adenomas, colitis cystica profunda) or submucosal lesions (eg, lipoma, leiomyoma) can have an endoscopic appearance similar to PI [27]. Endoscopic mucosal biopsies can rapidly deflate some cysts. PI can definitively be differentiated from these lesions by abdominal computed tomography scan. (See 'Endoscopic findings' above and 'Abdominal CT scan' above.)

MANAGEMENT

Emergent management — Patients with pneumatosis intestinalis (PI) and any **one** of the following should undergo emergent exploratory laparotomy (algorithm 1) [5,27].

- Signs of peritonitis on abdominal exam (eg, abdominal rigidity, rebound tenderness, and/or pain that worsens when the examiner lightly bumps the stretcher)
- Metabolic acidosis (arterial pH <7.3, bicarbonate [$HCO_{3]}$ <20 mmol/L)
- Lactate > 2.0 mmol/L
- Portal venous gas

Pneumoperitoneum caused by PI may be managed medically if there are no clinical features suggesting an underlying acute abdominal emergency [17]. (See 'Abdominal CT scan' above.)

Non-emergent management

Overview — In patients who do not require emergent exploratory laparotomy, management is based upon the severity of symptoms (algorithm 1). Mild symptoms include abdominal pain that does not interfere with functioning and the ability to perform activities of daily living.

Moderate to severe symptoms include abdominal pain or bleeding that interfere with functioning and the ability to perform activities of daily living.

Asymptomatic patients and those with mild symptoms can be managed as outpatients. Patients with moderate to severe symptoms require hospitalization.

- The underlying cause of PI should be treated in all patients, regardless of the presence of symptoms.
- Asymptomatic patients do not require additional therapy. Intramural gas cysts usually resolve spontaneously over time [18].
- For patients with mild symptoms, we use a combination of antibiotics and an elemental diet. (See 'Antibiotics' below and 'Elemental diet' below.)
- For patients with moderate to severe symptoms, we suggest a combination of antibiotics, an elemental diet, and inhalation oxygen therapy. For patients with continued symptoms despite a 10-day trial of inhalational oxygen therapy, we use hyperbaric oxygen therapy for three days. (See 'Oxygen' below.)
- We reserve surgery for patients who remain symptomatic despite medical therapy or who develop complications from PI (eg, bowel obstruction, perforation, or peritonitis) [43]. (See 'Surgery' below.)

Follow-up abdominal imaging (eg, abdominal computed tomography scan) should be performed once symptoms have resolved or every one to three months until there is radiographic resolution.

While several approaches have been used to treat patients with symptomatic PI, the evidence to support their efficacy is limited to small observational studies and these interventions have not been directly compared. Furthermore, recurrence within 18 months has been described in 30 to 40 percent of patients regardless of the choice of medical therapy [3,14,44-47].

Treatment of the underlying etiology — All patients should receive therapy directed against the underlying etiology of PI. Discontinuation of the culprit medication (eg, alpha-glucosidase inhibitors, molecular targeted therapy) may result in resolution of PI [23,48]. In one case series, in which 48 patients with PI or intestinal perforation following targeted molecular cancer chemotherapy with bevacizumab and sunitinib, PI resolved in all patients with discontinuation of targeted therapy [49]. Reinstitution of the therapy was associated with recurrence in the majority of patients in whom molecular targeted therapy was restarted after initial resolution.

Antibiotics — Antibiotics should be continued until there is clinical and radiographic resolution of PI or for up to three months [14,47]. We typically use metronidazole (500 mg orally three times daily) [14,47,50]. Patients should be informed of the potential adverse reactions associated with metronidazole (eg, nausea, vomiting, diarrhea, and neuropathy) and the need to avoid alcohol. For patients who cannot tolerate metronidazole, other antibiotics such as ampicillin, tetracycline, and vancomycin have been used successfully. Retreatment with a prolonged course has been effective in patients who relapsed following discontinuation [47].

Elemental diet — Resolution of PI has been reported in patients treated with an elemental diet, presumably by alteration of the colonic microflora [15,51]. The elemental diet should be continued for two weeks. In one series of two patients treated with pneumatosis refractory to medical therapy, symptoms resolved within four days of commencing the diet, and a colonoscopy at two weeks confirmed complete resolution of the cysts. Introduction of high-residue foods was associated with recurrence in one patient but resolved by restarting the elemental diet. (See "Nutrition support in critically ill patients: Enteral nutrition".)

Oxygen — Inhalational oxygen (fraction of inspired oxygen [FiO_2] of 55 to 75 percent to achieve a partial pressure of oxygen [PaO_2] of 200 to 350 mmHg for 4 to 10 days) or hyperbaric oxygen therapy (2.5 atmospheres for 2.5 hours on two or three consecutive days) have been used to treat PI [18]. However, the optimal amount and duration of oxygen therapy required for cyst deflation is unknown and treatment is associated with the risk of oxygen toxicity [3]. Short duration of treatment with hyperbaric oxygen therapy may help to avoid the pulmonary toxicity associated with prolonged high flow oxygen use [52]. (See "Adverse effects of supplemental oxygen".)

Inhalational and hyperbaric oxygen therapy have been associated with resolution of PI in several case reports [44,53-57]. The rationale for the use of oxygen therapy is twofold. First, oxygen is toxic to the anaerobic intestinal bacteria that contribute to gas cyst formation. Second, the contents of the gas cysts are primarily non-oxygen gases [11,12]. High concentrations of delivered oxygen increase the partial pressure of oxygen in the venous blood, and decrease the partial pressure of non-oxygen gases, such as nitrogen, creating a diffusion gradient across the cystic wall favoring the exit of gas from the cysts [10,20].

Surgery — Surgery should be reserved for patients with PI who remain symptomatic despite medical therapy or who develop complications from PI such as bowel obstruction, perforation, peritonitis, and necrotic bowel [43]. Although surgery can be effective, worsening of PI after surgery has also been reported [18].

Endoscopic therapy — Patients who develop obstructive symptoms from PI may not be candidates for surgery because of high perioperative risk. Successful relief of obstruction in such patients has been described using endoscopic puncture and sclerotherapy of the cysts [58].

SUMMARY AND RECOMMENDATIONS

- **Epidemiology** – Pneumatosis intestinalis (PI) may be an incidental finding in an asymptomatic patient that spontaneously resolves or may be seen in conditions associated with intestinal necrosis and indicate the need for emergency surgery. Adults are typically diagnosed in the fifth to eighth decade. (See 'Introduction' above and 'Etiology and pathogenesis' above.)

- **Etiology and pathogenesis** – PI can be seen in newborn infants and adults. PI is idiopathic (15 percent) or secondary (85 percent) to a wide variety of gastrointestinal and non-gastrointestinal illnesses. The pathogenesis of PI is likely multifactorial due to mechanical, bacterial, and biochemical causes (table 1). (See 'Epidemiology' above and 'Etiology and pathogenesis' above.)

- **Pathology** – PI can affect the small intestine, colon, or both. Cysts may be confined to the mucosa, submucosa, or subserosa, or involve all three layers. Subserosal cysts are more commonly seen in small intestinal pneumatosis, while submucosal cysts are more commonly seen in colonic pneumatosis. (See 'Pathology' above.)

- **Clinical features** – Most patients with PI are asymptomatic and probably never come to clinical attention. Patients who come to clinical attention present with symptoms related to either the presence of PI such as abdominal pain, obstruction, or bleeding, or due to the underlying disorder associated with PI (table 2).

- **Evaluation** – Assessment of patients with suspected PI should include a history, physical examination, laboratory studies, and abdominal imaging. We perform a contrast-enhanced abdominal computed tomography scan to establish the diagnosis, determine the underlying etiology, and diagnose associated complications. (See 'Evaluation' above.)

- **Indications for emergent management** – Emergent surgical exploration is indicated for patients with PI and any **one** of the following (algorithm 1) (see 'Overview' above):

  - Signs of peritonitis on abdominal exam (eg, abdominal rigidity, rebound tenderness, and/or pain that worsens when the examiner lightly bumps the stretcher).

- Metabolic acidosis (arterial pH <7.3, bicarbonate [HCO_3] <20 mmol/L).
- Lactate > 2.0 mmol/L.
- Portal venous gas.

- **Non-emergent management based on disease severity** – In patients who do not require emergent exploratory laparotomy, management is based upon the severity of symptoms (algorithm 1). The underlying cause of PI should be treated in all patients, regardless of the presence of symptoms.
  - **Patients without symptoms** – Asymptomatic patients do not require any additional therapy.
  - **Patients with mild symptoms** – For patients with mild symptoms who can be managed as outpatients, we suggest a combination of antibiotics and an elemental diet (**Grade 2C**). Follow-up imaging should be performed once symptoms have resolved or every one to three months until there is radiographic resolution. (See 'Antibiotics' above and 'Elemental diet' above.)
  - **Patients with moderate to severe symptoms** – For patients with moderate to severe symptoms who require hospitalization, we suggest a combination of antibiotics, an elemental diet, and oxygen therapy (**Grade 2C**). We reserve surgery for patients with PI who remain symptomatic despite medical therapy or who develop complications from PI such as bowel obstruction, perforation, peritonitis, and necrotic bowel. (See 'Antibiotics' above and 'Elemental diet' above and 'Oxygen' above.)

ACKNOWLEDGMENT — The UpToDate editorial staff acknowledges Paul Rutgeerts, MD (deceased), who contributed to earlier versions of this topic review.

REFERENCES

1. Cordum NR, Dixon A, Campbell DR. Gastroduodenal pneumatosis: endoscopic and histological findings. Am J Gastroenterol 1997; 92:692.
2. Sachse RE, Burke GW 3rd, Jonas M, et al. Benign pneumatosis intestinalis with subcutaneous emphysema in a liver transplant recipient. Am J Gastroenterol 1990; 85:876.
3. Heng Y, Schuffler MD, Haggitt RC, Rohrmann CA. Pneumatosis intestinalis: a review. Am J Gastroenterol 1995; 90:1747.

4. KOSS LG. Abdominal gas cysts (pneumatosis cystoides intestinorum hominis); an analysis with a report of a case and a critical review of the literature. AMA Arch Pathol 1952; 53:523.
5. Knechtle SJ, Davidoff AM, Rice RP. Pneumatosis intestinalis. Surgical management and clinical outcome. Ann Surg 1990; 212:160.
6. Pieterse AS, Leong AS, Rowland R. The mucosal changes and pathogenesis of pneumatosis cystoides intestinalis. Hum Pathol 1985; 16:683.
7. Pear BL. Pneumatosis intestinalis: a review. Radiology 1998; 207:13.
8. KAY-BUTLER JJ. Interstitial emphysema of the caecum. Gut 1962; 3:267.
9. KEYTING WS, MCCARVER RR, KOVARIK JL, DAYWITT AL. Pneumatosis intestinalis: a new concept. Radiology 1961; 76:733.
10. Davila AD, Willenbucher RF. Other diseases of the colon and rectum. In: Sleisenger & Fordtran's Gastrointestinal and Liver Disease, 6th ed, Feldman M, Scharschmidt B, Sleisenger M (Eds), WB Saunders Company, Philadelphia 1998. p.1979.
11. Read NW, Al-Janabi MN, Cann PA. Is raised breath hydrogen related to the pathogenesis of pneumatosis coli? Gut 1984; 25:839.
12. Florin TH, Hills BA. Does counterperfusion supersaturation cause gas cysts in pneumatosis cystoides coli, and can breathing heliox reduce them? Lancet 1995; 345:1220.
13. Yale CE, Balish E, Wu JP. The bacterial etiology of pneumatosis cystoides intestinalis. Arch Surg 1974; 109:89.
14. Ellis BW. Symptomatic treatment of primary pneumatosis coli with metronidazole. Br Med J 1980; 280:763.
15. van der Linden W, Marsell R. Pneumatosis cystoides coli associated with high H2 excretion. Treatment with an elemental diet. Scand J Gastroenterol 1979; 14:173.
16. Mehta SN, Friedman G, Fried GM, Mayrand S. Pneumatosis cystoides intestinalis: laparoscopic features. Am J Gastroenterol 1996; 91:2610.
17. Hoover EL, Cole GD, Mitchell LS, et al. Avoiding laparotomy in nonsurgical pneumoperitoneum. Am J Surg 1992; 164:99.
18. Sartor RB, Murphy ME, Rydzak E. Miscellaneous inflammatory and structural disorders of the colon. In: Textbook of Gastroenterology, 3rd ed, Yamada T, Alpers D, Laine L, et al (Eds), Lippincott Williams & Wilkins, Philadelphia 1999. Vol 1877.
19. Christl SU, Gibson GR, Murgatroyd PR, et al. Impaired hydrogen metabolism in pneumatosis cystoides intestinalis. Gastroenterology 1993; 104:392.

20. Levitt MD, Olsson S. Pneumatosis cystoides intestinalis and high breath H2 excretion: insights into the role of H2 in this condition. Gastroenterology 1995; 108:1560.

21. Hisamoto A, Mizushima T, Sato K, et al. Pneumatosis cystoides intestinalis after alpha-glucosidase inhibitor treatment in a patient with interstitial pneumonitis. Intern Med 2006; 45:73.

22. Hayakawa T, Yoneshima M, Abe T, Nomura G. Pneumatosis cystoides intestinalis after treatment with an alpha-glucosidase inhibitor. Diabetes Care 1999; 22:366.

23. Yanaru R, Hizawa K, Nakamura S, et al. Regression of pneumatosis cystoides intestinalis after discontinuing of alpha-glucosidase inhibitor administration. J Clin Gastroenterol 2002; 35:204.

24. Azami Y. Paralytic ileus accompanied by pneumatosis cystoides intestinalis after acarbose treatment in an elderly diabetic patient with a history of heavy intake of maltitol. Intern Med 2000; 39:826.

25. Jamart J. Pneumatosis cystoides intestinalis. A statistical study of 919 cases. Acta Hepatogastroenterol (Stuttg) 1979; 26:419.

26. Galandiuk S, Fazio VW. Pneumatosis cystoides intestinalis. A review of the literature. Dis Colon Rectum 1986; 29:358.

27. Höer J, Truong S, Virnich N, et al. Pneumatosis cystoides intestinalis: confirmation of diagnosis by endoscopic puncture a review of pathogenesis, associated disease and therapy and a new theory of cyst formation. Endoscopy 1998; 30:793.

28. Gelman SF, Brandt LJ. Pneumatosis intestinalis and AIDS: a case report and review of the literature. Am J Gastroenterol 1998; 93:646.

29. Kreiss C, Forohar F, Smithline AE, Brandt LJ. Pneumatosis intestinalis complicating C. difficile pseudomembranous colitis. Am J Gastroenterol 1999; 94:2560.

30. Sharma PG, Rajderkar DA, Sistrom CL, et al. Bubbles in the belly: How well do radiology trainees recognize pneumatosis in pediatric patients on plain film? Br J Radiol 2022; 95:20211101.

31. Vernacchia FS, Jeffrey RB, Laing FC, Wing VW. Sonographic recognition of pneumatosis intestinalis. AJR Am J Roentgenol 1985; 145:51.

32. Ribaldone DG, Bruno M, Gaia S, et al. Endoscopic ultrasound to diagnose pneumatosis cystoides intestinalis (with video). Endosc Ultrasound 2017; 6:416.

33. Varano VJ, Bonanno CA. Colonoscopic findings in Pneumatosis cystoides intestinalis. Am J Gastroenterol 1973; 59:353.

34. Rabushka LS, Kuhlman JE. Pneumatosis intestinalis. Appearance on MR examination. Clin Imaging 1994; 18:258.

35. Chaput U, Ducrotté P, Denis P, Nouveau J. Pneumatosis cystoides intestinalis: an unusual cause of distal constipation. Gastroenterol Clin Biol 2010; 34:502.

36. Tobias R, Coleman S, Helman CA. Pneumatosis coli simulating hepatomegaly. Am J Gastroenterol 1985; 80:146.

37. Ihara E, Harada N, Motomura S, Chijiiwa Y. A new approach to Pneumatosis cystoides intestinalis by target air-enema CT. Am J Gastroenterol 1998; 93:1163.

38. John A, Dickey K, Fenwick J, et al. Pneumatosis intestinalis in patients with Crohn's disease. Dig Dis Sci 1992; 37:813.

39. Kernagis LY, Levine MS, Jacobs JE. Pneumatosis intestinalis in patients with ischemia: correlation of CT findings with viability of the bowel. AJR Am J Roentgenol 2003; 180:733.

40. Wayne E, Ough M, Wu A, et al. Management algorithm for pneumatosis intestinalis and portal venous gas: treatment and outcome of 88 consecutive cases. J Gastrointest Surg 2010; 14:437.

41. Olson DE, Kim YW, Ying J, Donnelly LF. CT predictors for differentiating benign and clinically worrisome pneumatosis intestinalis in children beyond the neonatal period. Radiology 2009; 253:513.

42. Graber SD, Sinz S, Turina M, Alkadhi H. Pneumatosis intestinalis in abdominal CT: predictors of short-term mortality in patients with clinical suspicion of mesenteric ischemia. Abdom Radiol (NY) 2022; 47:1625.

43. Greenstein AJ, Nguyen SQ, Berlin A, et al. Pneumatosis intestinalis in adults: management, surgical indications, and risk factors for mortality. J Gastrointest Surg 2007; 11:1268.

44. Holt S, Gilmour HM, Buist TA, et al. High flow oxygen therapy for pneumatosis coli. Gut 1979; 20:493.

45. Grieve DA, Unsworth IP. Pneumatosis cystoides intestinalis: an experience with hyperbaric oxygen treatment. Aust N Z J Surg 1991; 61:423.

46. van der Linden W. Letter: Reappearance of intestinal gas cysts after oxygen treatment. Lancet 1974; 2:1388.

47. Tak PP, Van Duinen CM, Bun P, et al. Pneumatosis cystoides intestinalis in intestinal pseudoobstruction. Resolution after therapy with metronidazole. Dig Dis Sci 1992; 37:949.

48. Poor A, Braman SS. Pneumatosis Intestinalis Associated with the Tyrosine Kinase Inhibitor Nintedanib. Lung 2018; 196:373.

49. Shinagare AB, Howard SA, Krajewski KM, et al. Pneumatosis intestinalis and bowel perforation associated with molecular targeted therapy: an emerging problem and the role of radiologists in its management. AJR Am J Roentgenol 2012; 199:1259.
50. Jauhonen P, Lehtola J, Karttunen T. Treatment of pneumatosis coli with metronidazole. Endoscopic follow-up of one case. Dis Colon Rectum 1987; 30:800.
51. Johnston BT, McFarland RJ. Elemental diet in the treatment of pneumatosis coli. Scand J Gastroenterol 1995; 30:1224.
52. Masterson JS, Fratkin LB, Osler TR, Trapp WG. Treatment of pneumatosis cystoides intestinalis with hyperbaric oxygen. Ann Surg 1978; 187:245.
53. Miralbés M, Hinojosa J, Alonso J, Berenguer J. Oxygen therapy in pneumatosis coli. What is the minimum oxygen requirement? Dis Colon Rectum 1983; 26:458.
54. Shemen LJ, Stern H, Sidlofsky S, Myers ED. Treatment of pneumatosis cystoides intestinalis with high FIO2: report of two cases. Dis Colon Rectum 1979; 22:245.
55. Wyatt AP. Prolonged symptomatic and radiological remission of colonic gas cysts after oxygen therapy. Br J Surg 1975; 62:837.
56. Forgacs P, Wright PH, Wyatt AP. Treatment of intestinal gas cysts by oxygen breathing. Lancet 1973; 1:579.
57. Simon NM, Nyman KE, Divertie MB, et al. Pneumatosis cystoides intestinalis. Treatment with oxygen via close-fitting mask. JAMA 1975; 231:1354.
58. Johansson K, Lindström E. Treatment of obstructive pneumatosis coli with endoscopic sclerotherapy: report of a case. Dis Colon Rectum 1991; 34:94.

Topic 2596 Version 21.0

GRAPHICS

Diseases associated with pneumatosis intestinalis

Intra-abdominal catastrophe:	**Endoscopic procedures:**
▪ Intestinal ischemia ▪ Intestinal infarction ▪ Intestinal perforation ▪ Intestinal obstruction ▪ Necrotizing enterocolitis ▪ Typhlitis	▪ Esophagogastroduodenoscopy ▪ Colonoscopy ▪ Sclerotherapy ▪ Endoscopic retrograde cholangiopancreatography (ERCP)
Mucosal disruption:	**Diseases affecting gastrointestinal motility:**
▪ Peptic ulcer disease ▪ Crohn disease ▪ Ulcerative colitis ▪ Feeding jejunostomy tube ▪ Caustic ingestions ▪ Ruptured diverticulum	▪ Diabetes ▪ Scleroderma ▪ Hirschprung disease ▪ Intestinal pseudo-obstruction ▪ Jejunoileal bypass ▪ Pyloric stenosis/obstruction
Infections:	**Immunological disturbances:**

▪ Clostridioides difficile
▪ Tuberculosis
▪ Whipples disease
▪ Coronavirus disease 2019 (COVID-19)
▪ AIDS entrocolitides
  • Cryptosporidium
  • Mycobacterium avium
  ○ Intracellulare
  • Cytomegalovirus (CMV)
	▪ AIDS
	▪ Steroids
	▪ Chemotherapy
	▪ Molecular targeted therapy
	▪ Lymphoproliferative disorders
	▪ Bone marrow transplantation
	▪ Solid organ transplantation
	▪ Graft versus host disease
	▪ Amyloidosis
	▪ Collagen vascular disease

Pulmonary disorders:	
▪ Chronic obstructive pulmonary disease ▪ Asthma ▪ Cystic fibrosis ▪ Mechanical ventilation	

Graphic 58197 Version 5.0

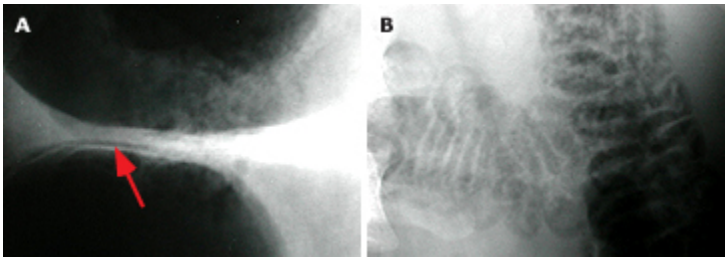
Symptoms of small and large bowel pneumatosis intestinalis

	Percent
Small Intestine	
1. Vomiting	60
2. Abdominal distention	59
3. Weight loss	55
4. Abdominal discomfort	53
5. Diarrhea	27
6. Anorexia	14
7. Constipation	12
Large Intestine	
1. Diarrhea	56
2. Hematochezia	50
3. Abdominal discomfort	32
4. Abdominal distention	28
5. Constipation	26
6. Weight loss	16
7. Tenesmus	10

Adapted from: Jamart J, Acta Hepato Gastroenterol 1979; 26:419.

Graphic 60778 Version 2.0

Pneumatosis intestinalis

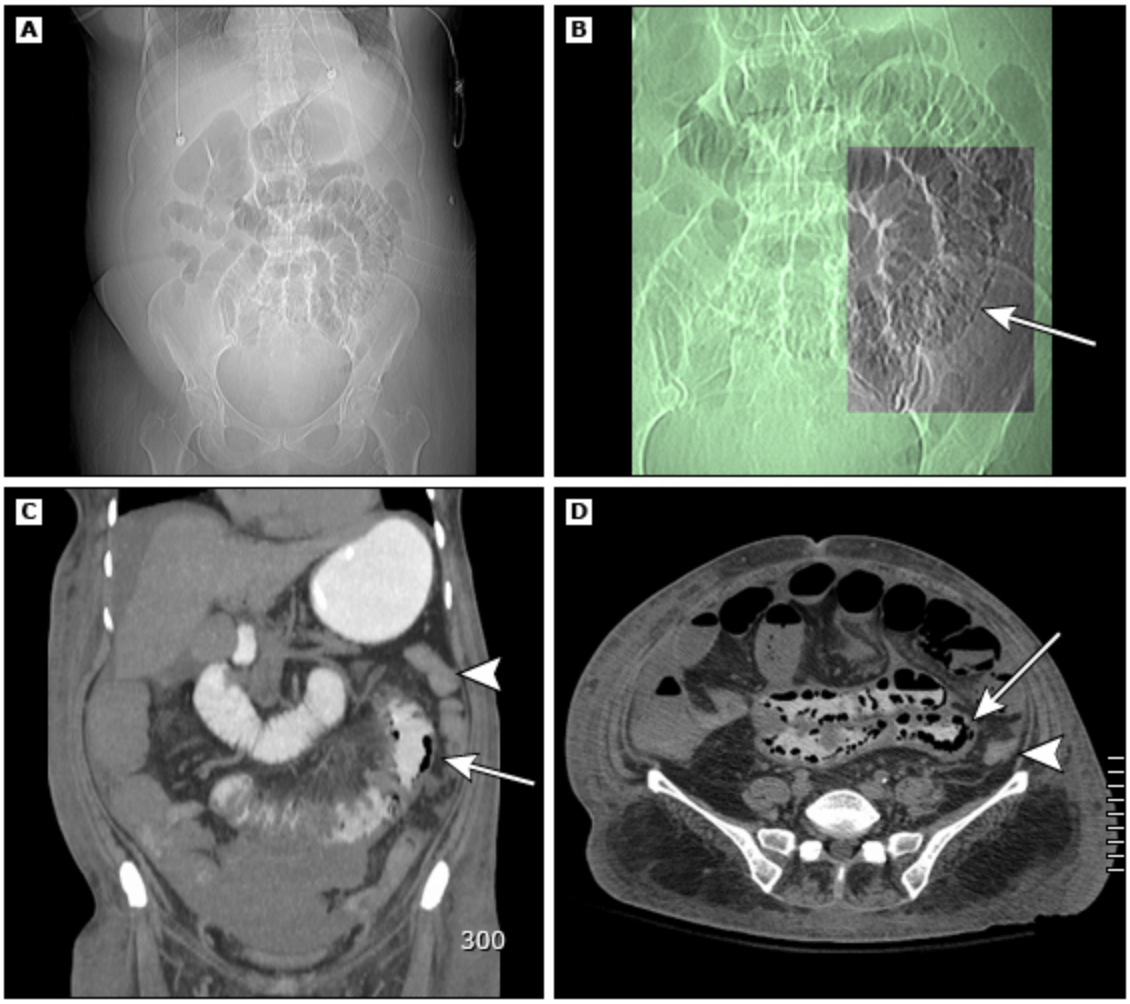


Panel A is a magnified image of a single fold in the colon demonstrating air within the wall of the colon, which appears as a thin linear lucency on this plain film (arrow). Panel B is a plain film from a 68-year-old male with ischemic bowel, demonstrating numerous linear collections of air within the bowel wall.

Courtesy of Jonathan B Kruskal, MD, PhD.

Graphic 72081 Version 2.0

CT scan of pneumatosis coli

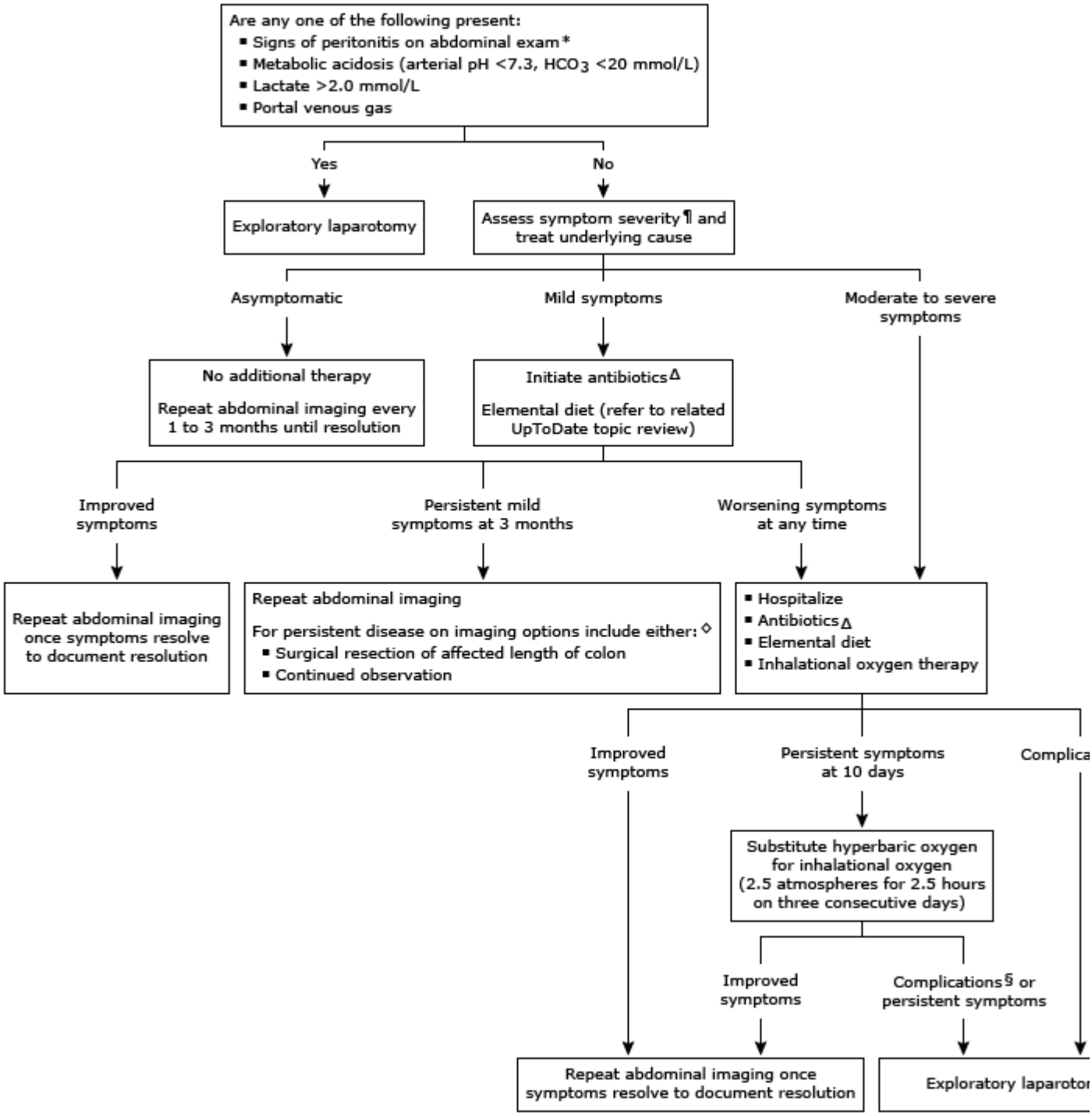


Computed tomography (CT) imaging of the abdomen is from a 55-year-old female who presented with acute abdominal pain and distension. Image A is a scout film of the abdomen and image B is a magnified view of the small bowel from the scout film. The images reveal air within the wall of the dilated loops of small-bowel, characteristic of pneumatosis intestinalis. The paucity of gas in the colon and rectum suggests SBO or severe ileus. Image C is coronal reformat through the abdomen confirming the accumulation of bubbles of air within the wall of the small bowel (white arrow). Image D is an axial image through the affected small bowel loop that shows more extensive accumulation of air within the wall of the thickened small bowel wall. Associated findings include decompressed large bowel (arrowheads), suggesting small bowel obstruction or severe ileus. In the appropriate clinical setting, the findings are highly suggestive of acute ischemia of the small bowel causing a severe ileus and functional obstruction. Less likely but also possible is small bowel obstruction with secondary ischemia.

CT: computed tomography; SBO: small bowel obstruction.

Graphic 58309 Version 3.0

Approach to the management of pneumatosis intestinalis in adults



* Abdominal rigidity, rebound tenderness, and/or pain that worsens when the examiner lightly bumps the stretcher.

¶ Mild symptoms include abdominal pain that does not interfere with functioning and the ability to perform activities of daily living. Moderate to severe symptoms include bleeding or abdominal pain that interferes wi functioning and the ability to perform activities of daily living.

Δ Antibiotic options include metronidazole, ampicillin, tetracycline, and vancomycin.

◇ Decision to recommend surgery depends on patient presentation, age, comorbidities, and extent of intest involved.

§ Complications include bowel obstruction, perforation, peritonitis, and necrotic bowel.

Graphic 110963 Version 1.0

Contributor Disclosures

Eric Goldberg, MD No relevant financial relationship(s) with ineligible companies to disclose. **J Thomas Lamont, MD** Equity Ownership/Stock Options: Allurion [Weight loss]. Consultant/Advisory Boards: Teledoc [Gastrointestinal diseases]. All of the relevant financial relationships listed have been mitigated. **Lawrence S Friedman, MD** Other Financial Interest: Elsevier [Gastroenterology]; McGraw-Hill [Gastroenterology]; Wiley [Gastroenterology]. All of the relevant financial relationships listed have been mitigated. **Shilpa Grover, MD, MPH, AGAF** No relevant financial relationship(s) with ineligible companies to disclose.

Contributor disclosures are reviewed for conflicts of interest by the editorial group. When found, these are addressed by vetting through a multi-level review process, and through requirements for references to be provided to support the content. Appropriately referenced content is required of all authors and must conform to UpToDate standards of evidence.

Conflict of interest policy

→