

# The Deceptive Nature of Pneumatosis Intestinalis: From Spontaneous Resolution to Bowel Ischemia

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**Background:** Pneumatosis intestinalis, a rare condition characterized by the presence of gas within the bowel wall, has an estimated incidence of 0.03% in the general population. Although the exact etiology of the condition remains uncertain, pneumatosis intestinalis is broadly classified as primary (idiopathic) or secondary (associated with an underlying condition), and the secondary form accounts for approximately 85% of cases. Secondary pneumatosis intestinalis has been associated with up to 60 potential causes and presents with a wide range of clinical manifestations, making thorough physical examination and imaging essential for appropriate management. Diagnosis is typically based on characteristic computed tomography (CT) findings.

**Case Report:** An 89-year-old female presented with shortness of breath, abdominal pain, nausea, vomiting, and diarrhea. CT imaging revealed extensive pneumatosis intestinalis in the small intestine. Despite the patient's stable hemodynamics and a mostly unremarkable physical examination, focal abdominal tenderness raised concern for bowel ischemia. The patient underwent an exploratory laparotomy that revealed patchy areas of pneumatosis intestinalis in an otherwise normal-appearing small bowel, with no evidence of ischemia or necrosis.

**Conclusion:** Because of the deceptive nature of pneumatosis intestinalis, the mere presence of the condition is not enough to justify surgery. Cases with benign causes can often be managed conservatively and resolve spontaneously, whereas concerning findings that may indicate a surgically treatable cause may require urgent intervention to reduce mortality.

**Keywords:** *Bowel, conservative treatment, ischemia, physiopathology, pneumatosis cystoides intestinalis, surgery*

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## INTRODUCTION

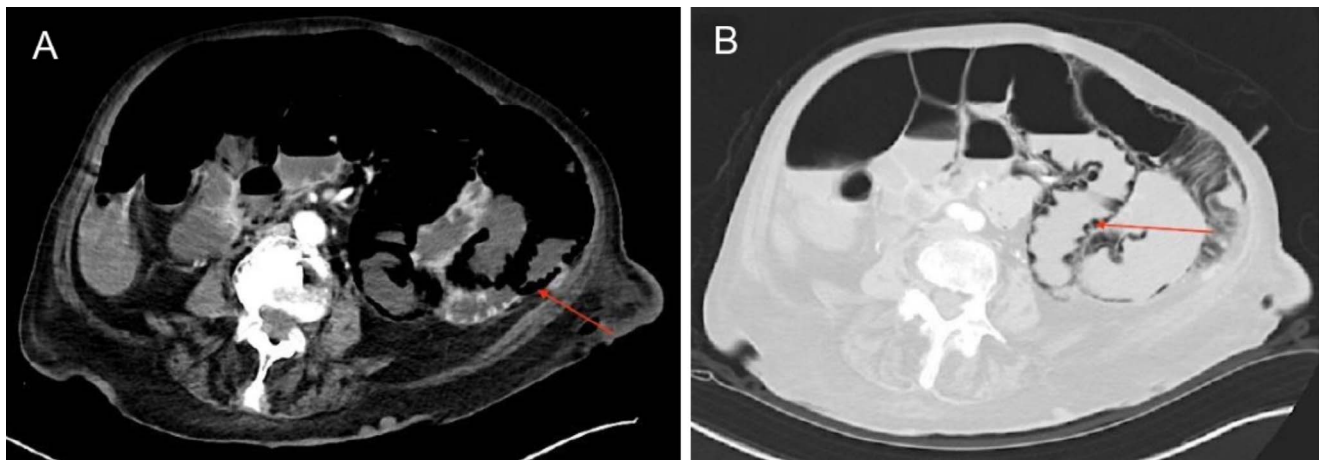
Pneumatosis intestinalis is an uncommon condition characterized by the presence of gas within the intestinal wall, typically affecting the mucosa and submucosa of the small and large intestines.<sup>1,2</sup> The gas formation is categorized as bubble-like (cysts in the intestinal wall) or band-like (continuous lines),<sup>3</sup> with the cysts located near blood vessels primarily on the antimesenteric aspect of the bowel.<sup>4</sup> While the band-like pattern is potentially indicative of bowel ischemia or portal venous gas, the bubble-like pattern is often harmless.<sup>3</sup>

Defined as a radiographic diagnosis by Lerner and Gazin in 1946,<sup>5</sup> pneumatosis intestinalis can occur at any age, depending on the underlying cause. Up to 60 causes—both benign and life-threatening etiologies—are associated with pneumatosis intestinalis,<sup>6</sup> which is classified as primary or secondary. The primary or idiopathic type (not associated with any other coexisting diseases) accounts for 15% of cases, while the secondary type, associated with a wide variety of underlying conditions, comprises the remaining 85% and includes life-threatening pathologies.<sup>7,8</sup> Mesenteric ischemia, bowel obstruction, and bowel necrosis

are common life-threatening causes. Pneumatosis intestinalis may also result from nonischemic and nonobstructive conditions, most of which are not linked to adverse outcomes.<sup>9</sup>

Patients with pneumatosis intestinalis may be asymptomatic or exhibit nonspecific symptoms such as abdominal pain, distention, and vomiting.<sup>10,11</sup> Given the potential for an ambiguous presentation, which can easily lead to a misdiagnosis, a thorough evaluation of the patient is essential, including a comprehensive history, identification of specific signs, and review of laboratory results.<sup>9,12,13</sup> Delays in surgical intervention can increase mortality.<sup>9</sup> Computed tomography (CT) scans are the gold standard imaging modality because they demonstrate greater sensitivity than plain radiography in differentiating between benign and concerning pneumatosis intestinalis.<sup>1,14</sup>

The priority in management is to determine whether the underlying pathology is benign or life-threatening. If a benign cause is suspected, conservative treatments are effective in 90% of cases.<sup>15</sup> Surgical management is reserved for instances in which conservative treatment fails, etiology is unclear, or a life-threatening condition is identified.<sup>15,16</sup>



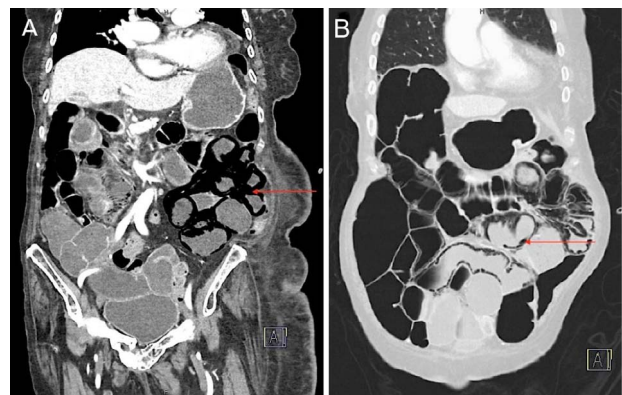
**Figure 1. Computed tomography with intravenous contrast (A, B) axial views of the abdomen and pelvis show diffuse pneumatosis intestinalis (arrows) in the small bowel.**

### CASE REPORT

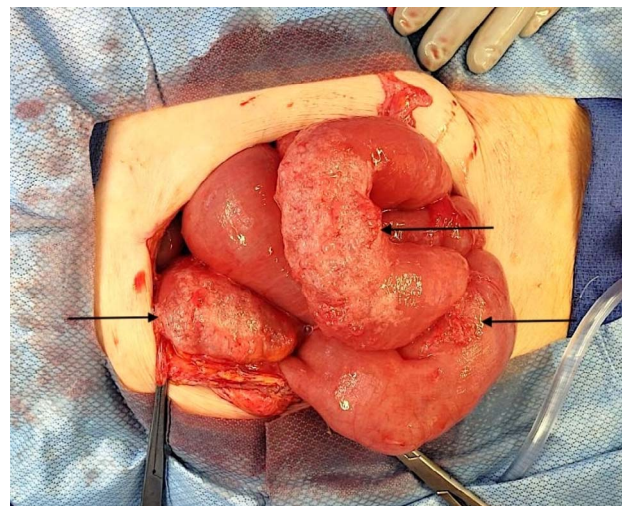
An 89-year-old female with a history of hypertension, gastroesophageal reflux disease, asthma, and duodenal ulcer perforation status post Graham patch repair was brought to the emergency department (ED) from her assisted living facility because plain radiographs showed left-sided thoracic infiltrate and possible pneumoperitoneum. Beginning 1 day prior, the patient complained of abdominal pain, nausea, vomiting, and nonbloody diarrhea. In the ED, she appeared mildly dyspneic, her abdomen was soft without peritoneal signs, and she had tenderness in the left lower quadrant. Initially, the patient was reportedly hypotensive, but she showed a sustained response to fluid administration. Complete blood workup revealed leukocytosis of  $16.23 \times 10^3/\mu\text{L}$  (reference range,  $4.30\text{-}10.50 \times 10^3/\mu\text{L}$ ) and hyponatremia of 127 mmol/L (reference range, 135-146 mmol/L); other laboratory findings were unremarkable, including a normal lactic acid level. Chest radiograph exhibited elevation of the left hemidiaphragm with loss of left basilar lung volume but no evidence of pneumoperitoneum. CT scan of the abdomen and pelvis with intravenous contrast showed distended loops of small bowel with pneumatosis intestinalis in the left abdomen but no evidence of pneumoperitoneum, bowel wall thickening, or portal venous gas (Figures 1 and 2).

Given the patient's overall stability and reassuring physical examination, a repeat CT scan with oral contrast was performed, which confirmed the previous findings and showed no evidence of mechanical bowel obstruction or contrast extravasation. However, because of the patient's initial hypotension, leukocytosis, hyponatremia, and persistent abdominal tenderness localized in the area where imaging showed the segment of small bowel with pneumatosis intestinalis, surgical exploration was recommended to rule out possible bowel ischemia.

Exploratory laparotomy identified large segments of the small bowel with pneumatosis intestinalis. Despite the presence of pneumatosis intestinalis, the bowel appeared pink and viable with no evidence of perforation, and no free fluid was found in the peritoneal cavity (Figure 3). Bowel resection was not performed, and the patient had an uncom-



**Figure 2. Computed tomography with intravenous contrast (A, B) coronal views of the abdomen and pelvis show diffuse pneumatosis intestinalis (arrows) in the small bowel.**



**Figure 3. Intraoperative image shows multiple segments of pneumatosis intestinalis (arrows) in the small bowel but no signs of necrosis.**

plicated postoperative course. She completed 7 days of empiric antibiotic therapy with metronidazole 500 mg every 8 hours and ciprofloxacin 400 mg every 12 hours, initially given intravenously and then switched to oral once the patient could tolerate diet.

Because of her baseline fragility and hyponatremia, the patient was not discharged until postoperative day 10. At her outpatient postoperative visit 2 weeks after discharge, she was doing well, and no concerns were identified during her physical examination.

## DISCUSSION

DuVernoy first described pneumatosis intestinalis in 1730.<sup>17</sup> In 1995, the incidence in the general population was reported to be 0.03%<sup>18</sup>; however, advanced imaging techniques have led to greater detection rates, with Morris et al reporting an incidence of 0.37% in 2008.<sup>19</sup> Pneumatosis intestinalis can occur at any age, with a higher incidence in older patients.<sup>9</sup> Although the entire gastrointestinal tract can be affected, the small bowel is the most common location (42%), followed by the colon (36%).<sup>20</sup>

Symptoms of pneumatosis intestinalis are not necessarily related to the presence or location of intramural gas in the gastrointestinal tract but rather to the underlying disorder.<sup>2,21</sup> Moreover, the extent of pneumatosis intestinalis does not correlate with the severity of symptoms or underlying disease.<sup>22</sup> Presentation is variable, ranging from asymptomatic incidental radiographic findings to a variety of gastrointestinal signs and symptoms such as abdominal distention, pain, nausea, and vomiting.<sup>10,11,21</sup> In a retrospective analysis of 919 cases of pneumatosis intestinalis, diarrhea, bloody stools, abdominal pain, constipation, weight loss, and tenesmus were the most common related symptoms.<sup>13</sup> Although pneumatosis intestinalis can arise from benign conditions that are neither obstructive nor ischemic,<sup>9</sup> life-threatening conditions typically involve bowel ischemia.<sup>6</sup> Delays in surgical intervention can result in high mortality rates, with reports showing that patients who undergo surgery within <6 hours tend to have a lower mortality rate.<sup>12</sup> Pneumatosis intestinalis in conjunction with a serum lactate concentration of  $\geq 2.0$  mmol/L, hypotension, metabolic acidosis, tachycardia, abdominal tenderness, and a thickened bowel wall has been associated with increased mortality.<sup>12</sup> Additionally, Hawn et al reported >80% mortality when serum lactate concentration was  $\geq 2.0$  mmol/L at the time of pneumatosis intestinalis diagnosis.<sup>23</sup>

While primary pneumatosis intestinalis is rare, a considerable number of diseases and disorders have been implicated as a potential cause.<sup>1,6</sup> Although the definite pathogenesis of pneumatosis intestinalis remains unclear, mechanical, bacterial, and pulmonary causes have emerged as possible explanations among the various theories of underlying mechanisms behind this pathology.<sup>9,10,11,24</sup>

The mechanical theory proposes that increased intraluminal pressure causes gas migration from the luminal space into the intestinal wall where the gas accumulates to form cysts.<sup>9</sup> The bacterial hypothesis suggests that air-producing bacteria penetrate the intestinal mucosal barrier, causing gas to become trapped within the submucosa and lymphatic channels.<sup>24</sup> This theory is supported by the observation that pneumatosis intestinalis has resolved with the use of metronidazole.<sup>15,25</sup> The pulmonary theory

attributes increased intraluminal pressure to the respiratory system.<sup>10,11</sup> Chronic lung conditions, such as chronic obstructive pulmonary disease, asthma, and interstitial pneumonia can cause alveolar rupture, allowing air to travel through the mediastinum into the perivascular spaces, ultimately reaching the intestinal wall.<sup>10,11,26</sup>

The diagnosis of pneumatosis intestinalis is generally based on findings from plain radiographs or CT scans that demonstrate extraluminal gas in the bowel wall.<sup>2,8</sup> CT is regarded as the more sensitive diagnostic tool because it can detect important secondary signs that may indicate an underlying cause of pneumatosis intestinalis, including bowel wall thickening, altered mucosal enhancement, abnormally dilated bowel, soft tissue stranding, portal venous gas, and ascites.<sup>27</sup> Imaging commonly also reveals features suggestive of free intraperitoneal gas.<sup>8</sup> Importantly, in the setting of pneumatosis intestinalis, pneumoperitoneum alone should not automatically suggest a perforated viscus, as free air can result from the perforation of individual gas-filled cysts.<sup>28</sup> CT imaging can help identify the distribution of gas and thereby assist in determining the cause of pneumatosis intestinalis.<sup>3</sup>

Useful laboratory tests include a complete blood count with differential, pH, serum bicarbonate, lactic acid, and serum amylase. Knechtle et al reported that the presence of metabolic acidosis, particularly lactic acidosis, and an elevated serum amylase level suggest ischemic bowel.<sup>21</sup> In the Knechtle et al study correlating laboratory results with ischemic bowel, pH <7.3, serum bicarbonate <20 mmol/L, lactic acid >2 mmol/L, and serum amylase >200 IU/L were predictive of necrotic bowel and were associated with lower survival rates compared to patients with normal values.<sup>21</sup>

Management should be guided by a combination of radiographic findings, physical examination, and a carefully obtained medical history, primarily focusing on determining if the underlying pathology of pneumatosis intestinalis is benign or life-threatening.<sup>9</sup> Patients with pneumatosis intestinalis on imaging and who present with abdominal pain, tenderness, diarrhea, fever, rectal bleeding, or hypotension are considered to have a severe clinical condition that requires surgery.<sup>21</sup> However, patients negative for abdominal pain, leukocytosis (white blood cell count <12 mm<sup>3</sup>), sepsis, or acidosis would benefit from medical support and observation.<sup>12</sup>

Therapy is tailored to the suspected etiology. For as many as 50% of patients, treatment is largely supportive and nonspecific.<sup>11</sup> While infectious causes usually require urgent intervention, pulmonary causes can be managed conservatively.<sup>29,30</sup> However, because the overall mortality rate in patients with pneumatosis intestinalis ranges from 22% to 50%, nonsurgical management should be approached with caution.<sup>8</sup>

In our case, although the patient was not peritonitic during the physical examination, she continued to have persistent abdominal tenderness in the area of the small bowel pneumatosis intestinalis identified on CT scan. Additionally, despite the absence of acidosis in her laboratory results, the patient arrived with hypotension and leukocytosis, concerns that led us to opt for surgical management to rule out bowel ischemia. The patient had no lung pathology or other conditions that could explain her physical examination findings, laboratory abnormalities, or imaging results.

Conservative management includes observation, hyperbaric oxygen, and antibiotic therapy.<sup>31</sup> Oxygen treatment is based on creating a high partial pressure of oxygen (PaO<sub>2</sub>) gradient that promotes gas outflow through the cyst wall. The cysts release the gas in them and refill with oxygen, which is subsequently metabolized, leading to resolution.<sup>31</sup> Antibiotic therapy primarily aims to reduce anaerobic bowel flora to mitigate gas production, and metronidazole is a commonly used agent.<sup>15,25</sup> Treatment typically continues until clinical and radiologic resolution is achieved.<sup>15</sup> On the other hand, surgery is the preferred treatment for patients experiencing severe abdominal pain and metabolic derangements such as acidosis, which more likely indicate a state of ischemic or threatened bowel.<sup>1,21,32</sup>

Although our patient ultimately did not benefit from surgical exploration, we believe that when a patient has an unclear clinical presentation, shows some signs of possible bowel ischemia, and does not have an obvious benign cause for pneumatosis intestinalis, a laparotomy may be beneficial.

## CONCLUSION

A thorough evaluation of patients with pneumatosis intestinalis should guide management, with consideration of the patient's history, physical examination, and diagnostic studies, including radiographic patterns. While cases with benign causes can often be treated conservatively, any concerning signs or symptoms that suggest a surgically correctable cause warrant consideration of urgent intervention to reduce the risk of mortality.

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