

PNEUMATOSIS INTESTINALIS

Definition: Air filled cysts within the wall of the intestine, when the colon is affected the term pneumatosis coli is applied.

Epidemiology: Uncommon disorder, true incidence unknown. The frequency is reportedly highest in the 6th decade. Equal frequency between men and women.

Etiology: There are 3 popular theories to explain pneumatosis and why it seen with so many conditions:

Mechanical: Gas enters thru a defect in the mucosa with or without increased luminal pressure (retching). This would explain-colitis, post-endoscopy, ulcer dz. However, the cysts have not been demonstrated to communicate w/ the mucosa making this less likely

Pulmonary: COPD, coughing leads to rupture of a bleb allowing air to dissect along vessels, the mediastinum, and retroperitoneum where it eventually situates itself in the subserosa of the GI tract. However, the absence of gas in the mediastinum and localized cysts argues against this.

Bacterial: Gas collections are by-products of bacterial metabolism. In lab animals-cysts can be reproduced by injecting clostridia into intestinal wall. In addition to local invasion, the bacteria may be producing large amounts of hydrogen from fermentation of carbohydrates. High H₂ concentration causes diffusion into the wall where it is followed by N₂, O₂, and CO₂ from the circulation- Breath hydrogen levels are increased in affected individuals. However, there is lack of evidence demonstrating bacteria within the cysts themselves.

Clinical Features: In the majority of cases it is an incidental finding on radiographs. Most common symptoms include: Diarrhea 70%, mucus discharge 70%, rectal bleeding 60%, and constipation 48%. Approximately 3 % of afflicted individuals experience a complication: pneumoperitoneum, volvulus, intestinal obstruction, intussusception, hemorrhage, or perforation. The clinical manifestations are usually a consequence of the underlying pathology. The pattern and extent does not correlate to the severity of the symptoms or the underlying disease.

The jejunum and ileum are the most frequently affected sites; the colon is affected in 6% of cases-with the left side of the colon being most frequently affected.

There are numerous conditions associated with pneumatosis:

Appendicitis	Emphysema/COPD
Crohn's	Collagen Vascular Diseases
UC	Transplantation
Diverticular Disease	AIDS
NEC	Steroid use
Pseudomembranous Colitis	Chemotherapy
Ileus	Sigmoid Volvulus
No comorbidity(20%-"primary pneumatosis")	

Diagnosis:

Plain films-demonstrate lucency within the wall of the GI tract. Findings are present in 2/3 of patients with pneumatosis.

Barium Enema has been used but the findings can be confused with polyposis as they have similar appearances.

Ultrasound can detect bright echogenic cysts in the bowel wall.

CT- Best modality and has a greater sensitivity than US or plain films.

Endoscopy- Macroscopic elevations of the mucosa. GI textbook recommends that all persons be scoped to rule out carcinoma or other causes. However, this should be done judiciously as biopsies of "polyps" have led to perforation

Treatment: Should be focused on the underlying illness.

1) In patients in whom surgery has no role very good results have been obtained with high flow oxygen-presumably by diffusion of hydrogen out of the cysts, oxygen into the cysts, then oxygen into the blood

2) Flagyl has been reported to be efficacious

3) Operative intervention?? Not on radiographic findings alone.

Usually reserved for non-responders of medical management, those with peritonitis, perforation, or abdominal sepsis.

Pneumoperitoneum has traditionally been associated with perforation. This is not necessarily the case with pneumatosis as pneumoperitoneum may represent a ruptured cyst(s). Patients with documented free air have been treated non-surgically and have similar, benign courses to those without this finding.

Gas in the portal system is associated with worse outcomes and often occurs with ischemic bowel. Prospective evaluations of pneumatosis have found 37% mortality associated with air in the portal venous system. In any case, air in the portal system is associated with worse outcomes but is not necessarily fatal.

4) Spontaneous regression in 50%

Take home points:

1) Usually not a surgical problem-However, experience appears to be largely anecdotal. Like any other patient need to look at the whole picture.

Sources:

1) Sleisenger and Fordtran's Gastrointestinal and Liver Disease 7th edition 2002 pgs 2307-2309

2) Shawn, D et al. "The Spectrum of Pneumatosis Intestinalis" Arch. Of Surgery Vol 138 Jan 2003. Pgs 68-75.

Medline search- "pneumatosis"

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