

# Pneumatosis intestinalis and volvulus: a rare association

## *Pneumatose intestinal e volvo: uma associação rara*

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### ABSTRACT

Pneumatosis intestinalis (PI) is a rare condition, especially when associated with volvulus; it is often misdiagnosed and inappropriately treated. We present the case of a 27 year-old woman suffering from an acute abdomen. An abdominal tomography was performed revealing Pneumatosis intestinalis. Once in the operating theatre sigmoid volvulus was diagnosed and Hartmann surgery performed. Histology showed intestinal ischemia. During the hospital stay, evolution was favourable. The authors present this case and a brief theoretical review, due to its rarity and clinical interest.

**Keywords:** Pneumatosis cystoides intestinalis; Abdomen, acute; Colon; Humans; Case reports

### RESUMO

A pneumatose intestinal (PI) é uma condição pouco frequente, sendo ainda mais rara em associação com volvo; sendo muitas vezes mal diagnosticada e tratada inapropriadamente. Apresentamos o caso de uma mulher de 27 anos com um quadro de abdómen agudo. Realizou TAC abdominal que demonstrou pneumatose intestinal. Intra-operatoriamente foi diagnosticado o volvo da sigmoideia e optado por cirurgia de Hartmann. O resultado anatomo-patológico da peça foi compatível com isquémia intestinal. Durante o internamento hospitalar, a doente evoluiu favoravelmente. Os autores apresentam este caso e uma breve revisão teórica, pela sua raridade e interesse clínico.

**Descritores:** Pneumatose cistoide intestinal; Abdome agudo; Colon; Humanos; Relatos de casos.

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### INTRODUCTION

Pneumatosis intestinalis (PI) is a rare disease that is characterized morphologically by the presence of multiple cysts gas located in the submucosa or subserosal of the intestinal wall<sup>(1-7)</sup>. It is a nonspecific signal, which can be found in a variety of clinical situations<sup>(1)</sup>. The primary form is rare (15% of cases) and generally affects the left colon. In 85% of cases, the PI is secondary to associated diseases (Table 1) as necrotizing enterocolitis in preterm infants, obstructive pulmonary disease or bowel ischemia, among others<sup>(1,3-8)</sup>. The secondary form mainly affects the small intestine and right colon<sup>(5)</sup>.

### CASE REPORT

The authors present the case of 22 year old woman, with medical history of epilepsy and psychomotor delay, suffering with diarrhea and prostration on the past 24 hours. On the clinical examination the patient was prostrated, with low grade fever (37.7°C), dehydrated, polypnea with good peripheral saturation, tachycardia, hypotensive (BP: 91/58mmHg), with a distended, painful abdomen, with abdominal guarding. Analytically, with increased inflammatory parameters (25 000 leukocytes, with neutrophilia and C-Reactive protein 20:57mg/dL). Urea 11mg/dL and creatinine 1.4mg/dL. An abdominal-pelvic computerized tomography (CT) was performed, that showed the presence of intestinal pneumatosis in the colon, with distension of the lumen with abundant liquid content, without significant thickening or fat densification adjacent to that; and a small amount of intraperitoneal fluid. Without other changes (Figure 1).

It was admitted an acute abdomen case, and the patient was taken to the emergency operating room. Intraoperatively, it was identified a colic redundancy, with exuberant sigmoid in apparent twist (volvulus), with multiple plaques of ischemia/necrosis involving the distal descending colon and the rectum-sigmoideia transition. An Hartmann's operation was decided. The anatomopathological report was consistent with extensive ischemic necrosis, from the mucosa and submucosa layers, with intense vascular congestion and signs of active peritonitis. The patient began empirical antibiotic therapy with piperacillin and tazobactam + metronidazole, with favorable evolution and improvement of the infection. She was discharged from the hospital, after fifteen days of treatment.

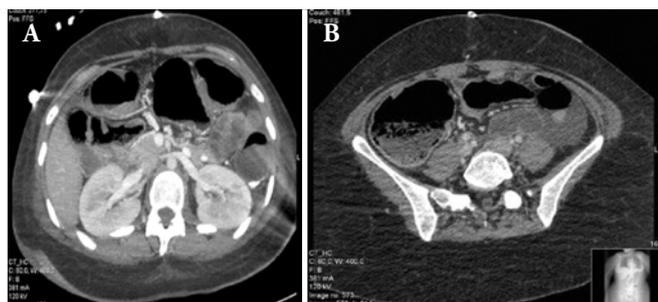
### DISCUSSION

The PI is most common in the 3rd and 4th decade of life, although it can occur at any age. There is no sex predominance<sup>(5)</sup>.

**Table 1.** Causes of secondary IP

Causes of secondary PI	
Pulmonary	Sistémicas
Chronic obstructive pulmonary disease	Systemic amyloidosis
Asthma	Collagen- vascular diseases
Cystic fibrosis	Autoimmune diseases
Chest trauma	“Donor- host” disease
Digestive	Organs transplantation
Mucosal disruption	Hemodialysis
Peptic ulcer	Polyarteritis nodosa
Caustic ingestion	Dermatomyositis
Bowel obstruction	Systemic lupus erythematosus
Rupture of diverticulum	Infectious
Abdominal trauma, volvulus	<i>Clostridium</i>
Surgery	Cytomegalovirus
Endoscopy	Human immunodeficiency virus
Mucosal injury by inflammation or ischemia	<i>Cryptosporidium</i>
Necrotizing enterocolitis	<i>Mycobacterium tuberculosis</i>
Ischemia or infarction	<i>Tropherima whippeli</i>
Appendicitis	Parasites
Crohn’s disease	Drugs
Ulcerative colitis	Steroids
	Immunosuppressive drugs
	Anesthesia with nitric oxide
	Lactulose/Sorbitol

Adapted<sup>(4)</sup>.



**Figure 1.** A and B) CT with intestinal pneumatosis.

First described in 1730 by DuVernoi, its etiopathogenesis is still unclear. There are several theories: mechanical (gas is forced to penetrate the intestinal wall through continuity solutions in the mucosa), lung (the released air from the rupture of alveoli slip by through the mediastinum, the aorta and mesenteric arteries to the intestine) and bacterial (gas-producing bacteria invade the wall or the resulting excess hydrogen from bacterial fermentation of carbohydrates in the lumen is absorbed and sequestered in the intestinal wall). The mechanical theory is the most widely accepted in the scientific community, however it seems that the increase in intraluminal pressure, high bacterial flora with consequent formation of intraluminal gas, and loss of mucosal integrity are conditions that interact in the formation

of PI<sup>(4,5,9)</sup>. The PI is often asymptomatic. Clinical manifestations include abdominal pain, intestinal obstruction, tenesmus, diarrhea and gastrointestinal bleeding<sup>(4)</sup>. Only in 3% of cases occurs complications including intestinal obstruction, intussusception, volvulus, bleeding and intestinal perforation<sup>(2,8,9)</sup>. A CT is the “gold standard” for the diagnosis of PI, but the simple abdominal radiography, ultrasound and endoscopy with biopsies collection help also to document the intestinal pneumatosis<sup>(4)</sup>. The finding of PI should be correlated with the patient’s symptoms and results of other complementary diagnostic tests<sup>(1)</sup>. Usually evolution is favorable. The treatment of symptomatic PI (primary or secondary) is medical, and consists on hyperbaric oxygen to increase the oxygen partial pressure<sup>(2,4,8)</sup>. Antibiotic therapy with metronidazole can be used, in view of the theory of bacterial PI.<sup>(1)</sup> The resolution of some secondary IP is based on adequate treatment of the underlying gastrointestinal disorder. Surgery is reserved for complicated cases with intestinal obstruction, volvulus, perforation, peritonitis and severe bleeding<sup>(1,2)</sup>. The association between PI and the sigmoid volvulus is rare. Some authors advocate the twist of the sigmoid as the initial event with subsequent ischemia of the mucosa and PI, while others suggest that PI as the cause of volvulus<sup>(10)</sup>. Given the rarity and variety of causes of PI, the great challenge of a doctor is to determine which findings of PI represent surgical indication. Although often benign, PI can present lethal complications, such as in this case. For this reason, the timely recognition of these complications can be “life-saving” and we must be allert to them<sup>(1,3)</sup>.

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