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Justin Bilello, MD
*Thomas Jefferson University*, justin.bilello@jefferson.edu

Amman Bhasin, MD
*Thomas Jefferson University*, amman.bhasin@jefferson.edu

Phoebe Chun, MD
*Thomas Jefferson University*, phoebe.chun@jefferson.edu

Aaron Martin, MD
*Thomas Jefferson University*, aaron.martin@jefferson.edu



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# End-Stage Chronic Intestinal Pseudo-Obstruction Resulting in Intestinal Pneumatosis

**Justin Bilello, MD[1], Amman Bhasin, MD[1], Phoebe Chun, MD[1], Aaron Martin MD[2]**

1. Internal Medicine Residency, Department of Medicine, Thomas Jefferson University Hospital, Philadelphia, PA

2. Division of Gastroenterology & Hepatology, Department of Medicine, Thomas Jefferson University Hospital, Philadelphia, PA

## ABSTRACT

Chronic intestinal pseudo-obstruction (CIPO) is a rare gastrointestinal motility disorder that presents with symptoms, physical exam, and imaging findings of mechanical bowel obstruction without an anatomical obstruction. Multiple etiologies, including enteric or extrinsic neuropathic dysfunction, myopathic dysfunction, or dysfunction of the interstitial cells of Cajal, cause CIPO's pathogenesis. The presentation of CIPO may be idiopathic or caused by underlying diseases. The most common presentation is abdominal pain, bloating, and distension. Here, we present a patient with an end-stage case of CIPO who failed medical therapy. Her distension progressed over a decade, requiring emergency surgery due to intestinal pneumatosis.

## INTRODUCTION

The enteric nervous system (ENS) has many responsibilities, including controlling gut musculature, secretory glands, and lymphatic blood vasculature. The delicate coordination of this system, colloquially referred to as the "brain-gut interaction," is paramount to the success of the gastrointestinal tract. Chronic intestinal pseudo-obstruction (CIPO) represents an intestinal motility failure that highlights the importance of a functional ENS and the impact on patient quality of life and healthcare costs.

CIPO is a rare disorder that presents on a spectrum of severity. The estimated prevalence of 0.80 per 100,00 with an incidence of 0.21 per 100,000.[1] CIPO is caused by enteric or extrinsic neuropathic dysfunction, myopathic dysfunction, or dysfunction of the interstitial cells of Cajal. The presentation of CIPO may be idiopathic or caused by underlying diseases. Sequelae of CIPO include anorexia, abdominal pain, small intestinal bacterial overgrowth, and malabsorption.[2]

Patients with CIPO present with clinical symptoms of intestinal obstruction without evidence of a mechanical obstruction; typically, patients suffer from bloating, abdominal pain, and vomiting. Radiologic imaging reveals air-fluid levels within distended bowel loops. Patients often require enteral or parental nutrition and opioid pain control with increased morbidity and mortality.[3] Management of CIPO necessitates a multidisciplinary team requiring supportive therapy, including nutritional and electrolyte supplementation and judicious pharmacotherapeutic medications.[4]

## CASE REPORT

A 40-year-old female with a history of AIGD (Autoimmune Gastric Dysmotility) complicated by CIPO presented to our medical center with several days of abdominal pain. On presentation, she described increased bloating and cramping abdominal pain. Vital signs and laboratory studies were within normal limits. She was admitted to the gastroenterology (GI) floor service for further management.

The patient has had long-standing abdominal bloating and pain dating back a decade. Full laboratory, endoscopic, and imaging workup was completed during initial GI outpatient visits to rule out irritable bowel disease, mechanical bowel obstruction, celiac disease, and infectious etiology. Ultimately, the patient was diagnosed with AIGD via a positive voltage-gated potassium channel antibody causing CIPO.

The patient rapidly became dependent on parental nutrition due to cachexia and the inability to tolerate oral nutrition. The patient was trialed on multiple medications to improve small bowel motility over several years, including metoclopramide, neostigmine, pyridostigmine, and prucalopride, with minimal response. Given the patient's bowel stasis, she also suffered from severe nausea and bloating. She was treated with multiple courses of rifaximin for presumed concomitant small intestinal bacterial overgrowth with little improvement. She ultimately required a venting gastrostomy tube placement, which was complicated by persistent peristomal leakage causing skin breakdown and significant pain. She was started on high-dose proton pump inhibitor therapy and octreotide with little improvement.

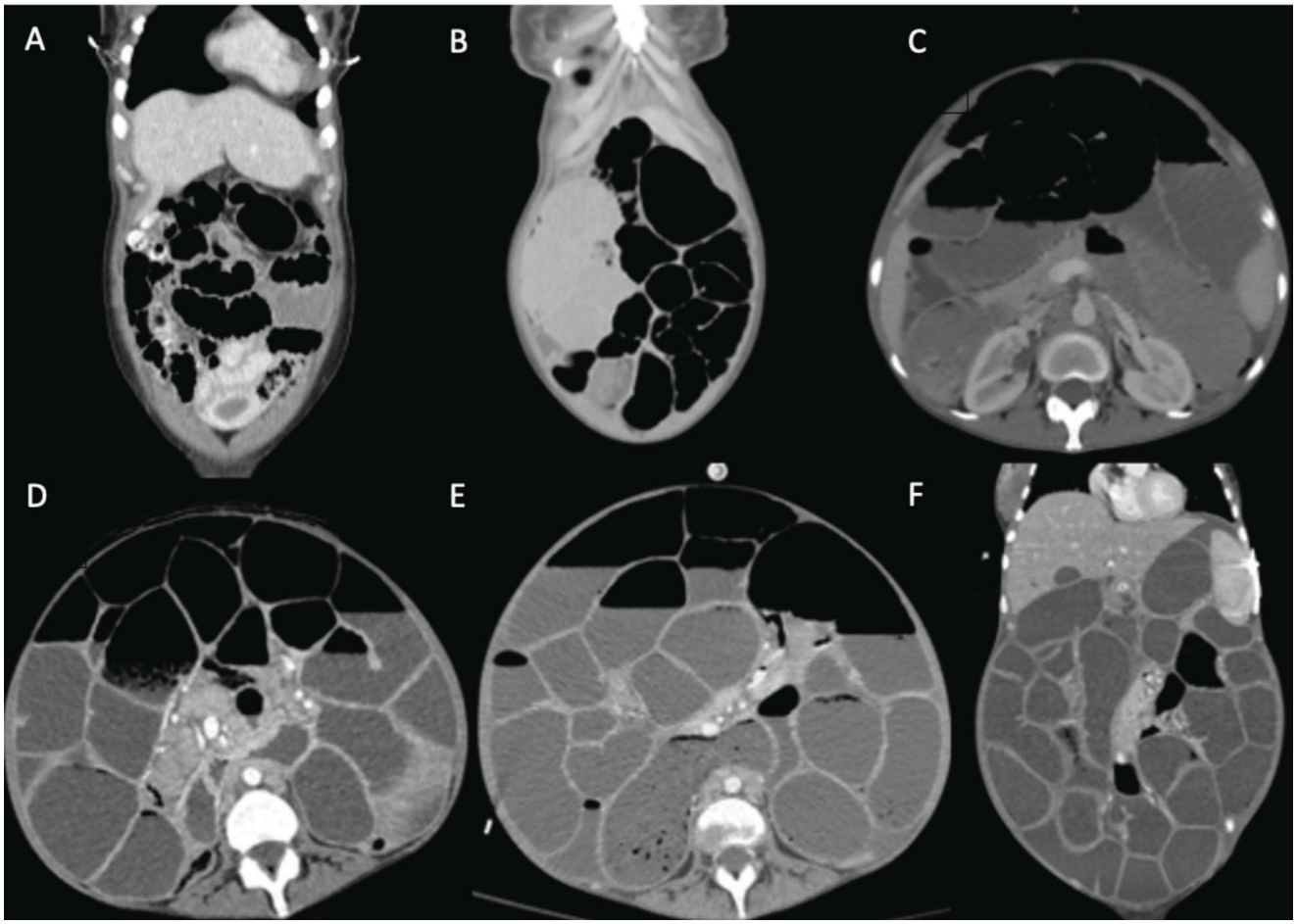


**Figure 1:** Computerized tomography (CT) imaging of the abdomen and pelvis demonstrating disease progression from 12/2013 to 2/2023. **A**) Coronal CT 12/16/2013 with unremarkable bowel gas pattern, **B**) Coronal CT 8/10/2014 with multiple distended small bowel loops and distension of the stomach, **C**) Axial CT 7/6/2017 with multiple loops of dilated large and small bowel with no discrete transition point, **D**) Axial CT 1/26/2023 with markedly diffuse small bowel dilation, **E** & **F**) Axial and coronal CT 2/26/2023 demonstrating severe small bowel obstruction with superimposed bowel ischemia with multiple foci of intramural, mesenteric venous and portal venous air.

Finally, given the failure of medical management, the patient was referred to another academic hospital for consideration of a small bowel transplant. Unfortunately, many complications and hospitalizations prevented the patient from undergoing transplant evaluation. She had numerous admissions from 2013 through 2023 for pain control, bacteremia, fungemia, gastrostomy site skin breakdown, and failure to thrive.

During the patient's most recent admission, she was evaluated emergently at the bedside for sinus tachycardia and abdominal pain, unresponsive to pain control. Given concern for bowel perforation, she was emergently sent for CT scan with intravenous contrast that revealed diffuse intestinal pneumatosis and portal venous air.

Given the patient's extreme distension, it was thought that her increased intraabdominal pressure was limiting blood supply to the small bowel. The patient was emergently taken to the surgical intensive care unit. After discussions between the gastroenterology team, the colorectal surgery team, and the patient, the decision was made to proceed to the operating room for abdominal exploration with possible resection of non-viable bowel and ileostomy creation.

While in the operating room, the small bowel was found to be massively distended (up to eight centimeters) with boggy, fibrinous exudate; however, the small bowel was completely viable without evidence of ischemia or need for resection. On inspection of the distal ileum, there was tapering to a normal caliber of the final ten centimeters. Ultimately, the decision was made to create a palliative loop ileostomy for possible distal decompression, given the patient's persistent abdominal pain, lactic acidosis, and imaging findings. The colon was inspected but also showed no signs of ischemia.

The small bowel was decompressed during the procedure, and eight liters of enteric contents were evacuated. An additional two liters of enteric contents were evacuated via a nasogastric tube. Ultimately, operative and pathology findings did not demonstrate ischemic bowel or mesenteric ischemia. Unfortunately, the patient's distension and abdominal pain were not significantly improved post-operatively despite the palliative surgical intervention.

## DISCUSSION

Though CIPO is a rare disease, it remains the leading cause of intestinal failure, with a significant burden on the healthcare system. While this patient's case may show a typical disease progression and representation of common complications over time, it also highlights intestinal pneumatosis, a rare complication of end-stage CIPO, in a patient who has failed all medical therapy. Further research and medical training devoted to improving diagnostic acumen, therapies, and quality of life in patients with CIPO is needed.

There are no diagnostic tests or pathognomonic findings unique to CIPO. The median time to diagnosis is eight years after symptom onset, and patients typically undergo multiple unnecessary surgeries and procedures in this interim.[2] During this time, patient quality of life is poor, with many dependent on parental nutrition, unsuccessful trials of expensive medications, and developing a dependence on narcotics for pain.[5] As a result, patients with CIPO have frequent hospital admissions, prolonged lengths of stay, and a high-cost burden in the healthcare system. A standardized diagnostic approach is needed to promptly identify underlying disease and pathology to alleviate patient suffering.[6]

The inception of gut transplantation in the 1990s has added another dimension to the management of CIPO to offer patients nutritional autonomy and improved quality of life.[7] Further research devoted to developing a multidisciplinary team approach, including surgeons, nutritionists, gastroenterologists, and pain management specialists, can improve quality of life and more rapidly identify transplant candidacy.

While current treatment aims to alleviate symptoms, new therapies targeting the ENS may be on the horizon as further advances within stem-cell therapy, pharmacology, and surgical techniques continue. These advancements are pivotal to avoiding poor outcomes, such as this patient's case.

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### Contact Information:

Justin Bilello, MD
Justin.Bilello@jefferson.edu
(484) 252- 6898


### Disclosures:

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