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# Development of Achalasia in a Patient With Chronic Intestinal Pseudo-Obstruction

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## ABSTRACT

Development of concurrent achalasia and chronic intestinal pseudo-obstruction (CIPO) is rare, although esophageal dysmotility is common in patients with CIPO and may suggest worse clinical outcomes. We present a case of a 63-year-old man with a 15-year history of CIPO who developed postprandial regurgitation, vomiting, and dysphagia and was diagnosed with achalasia through radiographic and endoscopic findings. This is only the third case in the reported literature that involves both conditions. Rather than representing 2 separate disorders, CIPO and achalasia may instead represent neurogenic variants of 1 underlying condition affecting the myenteric plexus.

## INTRODUCTION

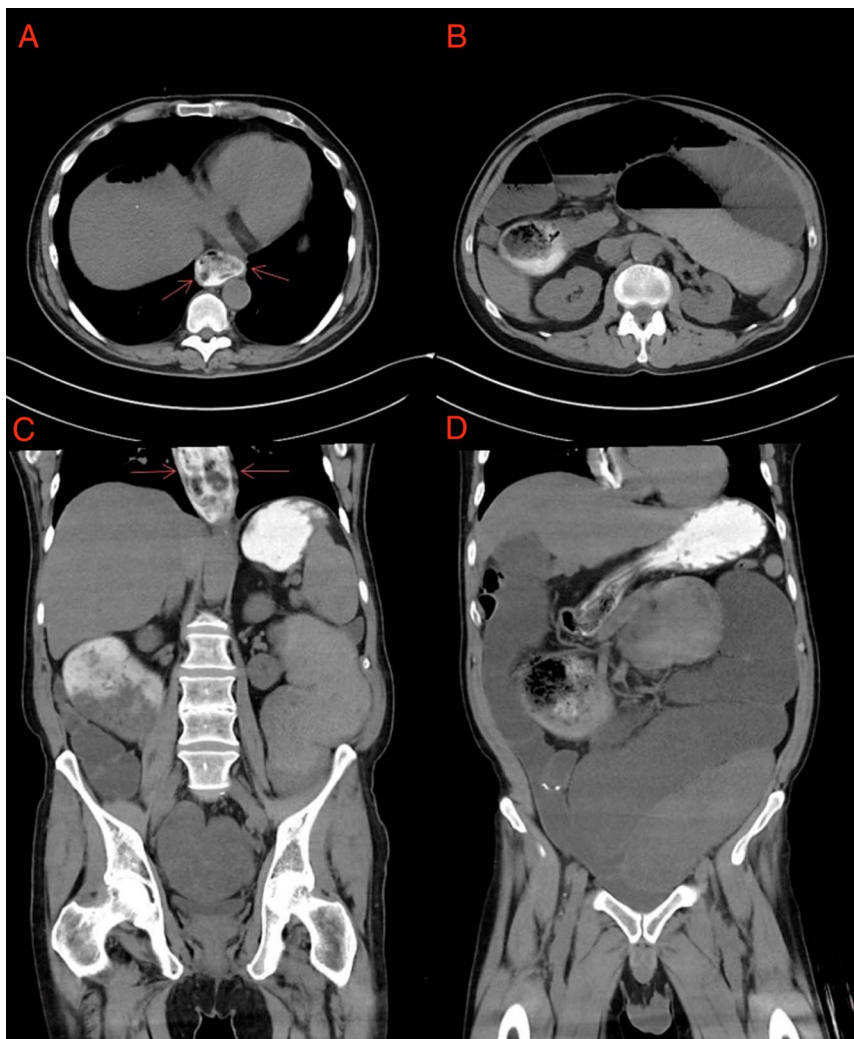
Chronic intestinal pseudo-obstruction (CIPO) is a rare gastrointestinal motility disorder characterized by signs and symptoms of bowel obstruction in the absence of any lesions occluding the intestinal lumen.<sup>1,2</sup> The disease may occur secondary to neuropathic or myopathic processes; however, the exact etiology is often unclear.<sup>3</sup> Achalasia is also a rare motility disorder of the esophagus but is characterized by impaired relaxation of the lower esophageal sphincter (LES) and absence of esophageal peristalsis.<sup>4</sup> The development of these 2 conditions is rare, with 2 previous case reports in the medical literature.

## CASE REPORT

A 63-year-old white man with a 15-year history of idiopathic CIPO presented for evaluation. His gastrointestinal history began 15 years earlier when he developed a cecal volvulus and underwent ileocecal resection. Postoperatively, he developed persistent abdominal pain with imaging concerning for bowel obstruction. He underwent 2 exploratory laparotomies, which showed no evidence of mechanical obstruction or transition point. Intestinal transit studies showed delayed gastric emptying, and small bowel transit, and follow-up computed tomography (CT) scans showed persistently severely dilated bowel loops. He was not taking opioid or anticholinergic medications. He had no history of type 2 diabetes, celiac disease, connective tissue disease, or other skeletal muscle disorders. A paraneoplastic antibody screen was negative.

Gastroduodenal manometry demonstrated an abnormal contractile pattern consistent with a neuropathic process, and he was diagnosed with CIPO. Consequently, despite trials of several motility agents, including metoclopramide, prucalopride, and pyridostigmine, he was dependent on total parental nutrition (TPN) for the last 10 years because of symptoms and poor oral intake. He had also been treated with rotating antibiotics for small intestinal bacterial overgrowth and omeprazole for gastroesophageal reflux disease. He was initially treated with omeprazole 40 mg twice daily for 12 weeks and subsequently decreased to 20 mg daily with symptom improvement.

During a follow-up visit, the patient described new symptoms of postprandial nausea with vomiting, food regurgitation, and solid food dysphagia. An abdominal CT scan showed dilation of the distal thoracic esophagus and retention of oral contrast with narrowing of the gastroesophageal junction. Other findings included chronically dilated small bowel to the level of the terminal



**Figure 1.** Abdominal computed tomography with oral contrast. (A and B) Axial view demonstrates a dilated, fluid-filled esophagus at the level of gastroesophageal junction and evidence of chronically dilated bowel. (C and D) Coronal views of the dilated esophagus and dilated bowel are also provided. Red arrows outline the dilated esophagus in both views.

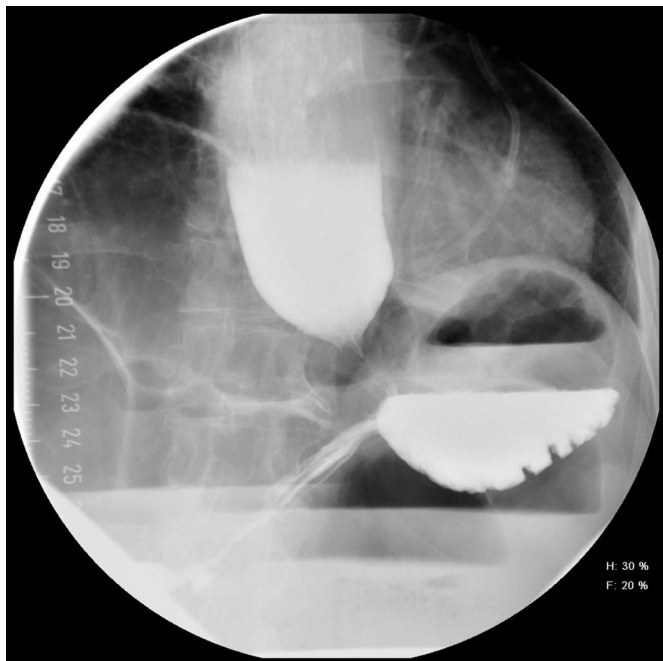
ileum, consistent with previous imaging and diagnosis of CIPO (Figure 1). A timed barium esophagram showed esophageal dilation with a bird beak appearance of the LES and a 5-cm fluid level of retained barium after 5 minutes (Figure 2). Esophagogastroduodenoscopy (EGD) showed a hypertonic LES with retention of saliva, mild esophagitis, absence of esophageal peristalsis, and resistance to endoscopic advancement into the stomach, concerning for achalasia (Figure 3). High-resolution esophageal manometry was attempted, but the study was prematurely aborted because of significant patient discomfort and inability to pass the catheter.

A repeat EGD was performed, which showed severe erosive esophagitis of the distal esophagus. Two mucosal tears were noted with the passage of the endoscope, so neither a manometry catheter (to later perform esophageal manometry) nor the endoluminal functional imaging probe was passed to limit further mucosal disruption. Based on clinical symptoms and findings seen on CT imaging, esophagram, and EGD, the patient was

diagnosed with achalasia and referred to surgery for myotomy. He was referred for a Heller myotomy based on local expertise and a lower suspicion for spastic type III achalasia, given the clinical history. Given CIPO and TPN dependence, it was felt that surgery would not improve his chances of tolerating a diet but rather reduce his symptoms of regurgitation, dysphagia, and risk of aspiration. A Heller myotomy with fundoplication was successfully performed with no major complications. One month after the procedure, the patient was seen in the clinic where he reported improved nausea and dysphagia.

## DISCUSSION

This is a case presentation of a patient with a long-standing history of CIPO and a new diagnosis of achalasia, the third described case of these 2 diagnoses in the medical literature.<sup>5,6</sup> Both previous cases describe patients who first developed achalasia and then developed CIPO several years later. Here, we describe the first case of a patient with long-standing CIPO and



**Figure 2.** Timed barium esophagram with retained contrast at the distal esophageal junction and classic “bird beak narrowing” consistent with achalasia.

subsequent diagnosis of achalasia. CIPO does not represent a single disease entity but rather encompasses a group of disorders, resulting in failure of gut motility.<sup>1</sup> Familial forms of CIPO have been reported, but most cases are believed to be sporadic.<sup>2</sup> Although CIPO may be secondary to known pathological conditions, the etiology is often idiopathic. Examples of secondary causes include paraneoplastic syndromes, radiation, connective tissue disorders (ie, Ehlers-Danlos syndrome), endocrinopathies (ie, hypothyroidism), autoimmune disease (ie, celiac disease), medication-induced (opioids), or infection (ie, Chagas disease).<sup>7</sup> With a full-thickness biopsy, CIPO can be classified into 3 categories depending on the histopathological abnormality: myopathy (smooth muscle cells), mesenchymopathy (interstitial cells of Cajal), and neuropathy (enteric nervous system).<sup>3</sup> The cause of CIPO in this case presentation was believed to be neuropathic.

Although CIPO and achalasia describe separate disease entities, they may represent manifestations of the same underlying pathologic process. In achalasia, failure of the LES to relax is hypothesized secondary to loss of inhibitory innervation of the esophageal myenteric plexus.<sup>8</sup> Similarly, CIPO may occur secondary to a neuropathic process affecting the myenteric plexus, and thus, these disorders may represent neurogenic variants of 1 underlying condition.<sup>6</sup> Furthermore, in a previous study of patients with gastrointestinal motility disorders, antiganglionic acetylcholine receptor antibodies were found in 21.4% of patients with achalasia and 50% of patients with CIPO. This suggests that antiganglionic acetylcholine receptor antibodies may mediate autonomic dysfunction and contribute to an immune-mediated mechanism underlying these 2 diseases.<sup>9</sup>



**Figure 3.** Esophagogastroduodenoscopy of the distal esophagus with evidence of narrowing at the lower esophageal sphincter, along with some evidence of esophagitis.

Although reports of overt achalasia and CIPO are rare, esophageal involvement with dysmotility is common in CIPO. In a retrospective study of 71 patients with CIPO who underwent esophageal manometry, 73% had abnormal motility patterns, including 68% with ineffective esophageal motility and 5% (4 patients) with vigorous achalasia. The 4 patients with achalasia were not further described. Esophageal motor disorders were associated with reduced survival, TPN requirement, and inability to maintain sufficient oral feeding.<sup>10</sup> Data remain limited on outcomes in patients with achalasia and CIPO. In conclusion, we describe a case presentation of a patient with CIPO who developed new esophageal symptoms and was subsequently diagnosed with achalasia. These 2 diseases may represent different manifestations of 1 underlying pathologic process. More research is required regarding the pathophysiological connection between achalasia and CIPO and the clinical outcomes in these patients.

## DISCLOSURES

Author contributions: J. Kwon wrote the manuscript and revised the manuscript for intellectual content. A. Koop and D. Francis revised the manuscript for intellectual content. D. Francis is the article guarantor.

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Informed consent was obtained for this case report.

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