Chronic intestinal pseudo-obstruction: Management

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INTRODUCTION

Pseudo-obstruction is a syndrome characterized by signs and symptoms of a mechanical obstruction of the small or large bowel in the absence of an anatomic lesion that obstructs the flow of intestinal contents. Pseudo-obstruction may be acute or chronic and is characterized by the presence of dilatation of the bowel on imaging. When there is evidence of chronic small intestinal motility disorder in the absence of bowel dilatation, the preferred term is chronic intestinal dysmotility.

This topic review will discuss the management of chronic intestinal pseudo-obstruction (CIPO). The etiology, clinical manifestations, and diagnosis of CIPO, acute colonic pseudo-obstruction (Ogilvie syndrome), chronic intestinal dysmotility, and slow transit constipation/colon inertia are discussed separately. (See "Chronic intestinal pseudo-obstruction: Etiology, clinical manifestations, and diagnosis" and "Acute colonic pseudo-obstruction (Ogilvie's syndrome)" and "Etiology and evaluation of chronic constipation in adults".)

INITIAL MANAGEMENT

Initial management of chronic intestinal pseudo-obstruction (CIPO) consists of dietary modification and treatment of the underlying disease. In patients with continued symptoms, pharmacologic therapy with prokinetic and antiemetics may be required. Patients should be

managed by a multidisciplinary team including a gastroenterologist, nutritionist, and a general, and in some cases a transplant, surgeon with experience in the treatment of CIPO [1,2].

General measures in all patients

Dietary modification — Early intervention with nutritional support is important, particularly for those who have had recurrent vomiting or reduced oral intake. Small meals consisting of liquid or homogenized foods are better tolerated than solids. Patients with CIPO frequently require supplemental nutritional support. Hypercaloric liquid formulations should be used in patients with low caloric intake. Enteral nutrition is typically used in patients with a neuropathic disorder and in whom the motility disorder is localized to the stomach and duodenum. Parenteral nutrition may be necessary for patients with severe dysmotility (usually myopathic pseudo-obstruction). (See 'Decompression and feeding' below and 'Parenteral nutrition' below.)

Treatment of the underlying disease — Treatment should be directed at the underlying disease. As an example, enzyme replacement therapy has been associated with alleviation of gastrointestinal manifestations in patients with Fabry disease. (See "Fabry disease: Clinical features and diagnosis" and "Fabry disease: Treatment and prognosis".)

Symptomatic management — Pharmacologic therapy is necessary for patients with severe symptoms or those who continue to have symptoms despite dietary modification. (See "Characteristics of antiemetic drugs".)

Prokinetics

- Prucalopride – 5HT4 receptor agonist, prucalopride, accelerates transit through the stomach, small bowel, and colon [3]. In a small randomized trial, prucalopride appeared to provide symptom relief in four of seven patients with CIPO [4]. In three patients with visceral myopathy and one with visceral neuropathy, 2 to 4 mg prucalopride (relative to placebo) significantly improved pain in three of four patients, nausea in two, vomiting in one, and bloating in four, whereas the frequency of bowel movements did not significantly change.

- Pyridostigmine – Pyridostigmine, an oral anticholinesterase, may be used in the chronic phase of CIPO. In small observational studies, pyridostigmine has demonstrated efficacy in the treatment of CIPO in adults and in children [5,6].

- **Other prokinetic agents** – We avoid the use of other prokinetics (eg, metoclopramide and erythromycin) for management of chronic symptoms of CIPO due to their side effects and tachyphylaxis associated with erythromycin.

Antiemetic agents — In patients with persistent symptoms despite the use of prokinetics, addition of an antiemetic medication may help to relieve symptoms of CIPO [7]. However, when coadministering antiemetics and prokinetics, it is important to consider potential metabolic drug interactions that may result in high blood levels resulting in drug toxicity. We treat patients with persistent symptoms despite prokinetics with an antiemetic medication (eg, promethazine 12.5 to 25 mg twice daily administered orally or as a rectal suppository or prochlorperazine) for symptom relief. The sedative effects of promethazine may be helpful when administered at bedtime. In patients who cannot tolerate promethazine, a 5-HT3 antagonist (eg, oral ondansetron 4 to 8 mg three times daily) may be used. However, 5-HT3 antagonists can delay colonic transit. We therefore reserve their use in patients who cannot tolerate promethazine or prochlorperazine. (See "Characteristics of antiemetic drugs".)

Additional therapies in selected patients

Patients with bacterial overgrowth — Small intestinal bacterial overgrowth (SIBO) is suspected in patients with CIPO who have flatulence, chronic diarrhea, steatorrhea, and vitamin deficiencies (eg, fat-soluble vitamins and vitamin B12) that may not be absorbed normally.

Antibiotics — Patients with suspected SIBO should be treated empirically with antibiotics. Breath tests for SIBO are limited by large variations in their performance and interpretation in patients with severe dysmotility [8]. Jejunal aspirate cultures are needed if steatorrhea does not respond to empiric antibiotics. Jejunal aspirate culture is the preferred test for diagnosing SIBO in patients with CIPO as breath tests can have a high false-negative rate in patients with motility disorders [9]. The presence of ≥10^5 aerobic colony forming units/mL in the jejunal aspirate is consistent with SIBO. (See "Small intestinal bacterial overgrowth: Management", section on 'Antibiotic therapy' and "Small intestinal bacterial overgrowth: Clinical manifestations and diagnosis", section on 'Diagnosis'.)

In patients with severe myopathic disease with recurrent SIBO, antibiotic prophylaxis may be needed. (See "Small intestinal bacterial overgrowth: Management", section on 'Antibiotic prophylaxis in selected patients'.)

Octreotide — We avoid the use of octreotide in patients with CIPO since it delays gastric emptying and small bowel transit of solids, which may be deleterious to patients with CIPO [10,11]. If it is to be used, it should only be administered before bedtime (at least two hours after the last meal).

Octreotide induces migrating motor complexes (MMCs), and limited evidence suggests that it may reduce the risk of bacterial overgrowth in patients with CIPO by moving food residue and possibly bacteria from the small intestine to the colon. In a series of five patients with

scleroderma, octreotide (50 mcg administered subcutaneously at bedtime) caused an average of 3.6 MMCs in three hours of small bowel recordings when there had previously been none [12]. There was a significant reduction in breath hydrogen excretion as compared with baseline and a reduction in symptoms of nausea, bloating, and abdominal pain and emesis. A similar benefit to intestinal motility with improvement in symptoms was observed in a few patients with idiopathic intestinal pseudo-obstruction who were treated with octreotide in combination with erythromycin [13]. However, symptomatic improvement with octreotide may reflect changes in visceral afferent function rather than an effect upon transit through the intestine [14,15].

Patients with paraneoplastic or inflammatory neuropathy — Immunosuppressive therapy should be reserved for patients with CIPO due to an underlying inflammatory neuropathy that is established by biopsy and in patients with a paraneoplastic neuropathy by the presence of antineuronal antibodies (anti-Hu) [16]. In addition to treatment of the underlying tumor, immune therapy with glucocorticoids (methylprednisolone or prednisone starting at 40 to 60 mg daily) may be needed. Rituximab and cyclophosphamide have also been used [17]. (See "Paraneoplastic syndromes affecting spinal cord, peripheral nerve, and muscle", section on 'Autonomic neuropathy'.)

Management of acute exacerbations — Patients with acute exacerbations of CIPO are typically hospitalized and require intravenous fluids. Medications that can exacerbate CIPO should be discontinued (eg, opiates, calcium channel blockers, medications with anticholinergic side effects). If the patient has received mu-opiates, a peripherally acting mu opioid receptor antagonist such as subcutaneous methylnaltrexone (0.15 mg/kg) may improve symptoms. (See "Prevention and management of side effects in patients receiving opioids for chronic pain", section on 'Methylnaltrexone'.)

Erythromycin — We administer intravenous erythromycin lactobionate at a dose of 3 mg/kg every eight hours for at least five to seven days. Intravenous erythromycin is effective during acute exacerbations of CIPO, acting at least in part by stimulation of the motilin receptors [18,19]. In patients with systemic sclerosis (scleroderma) who have impaired gastric motility, erythromycin can improve gastric emptying [20]. However, we do not use erythromycin for chronic therapy as there is limited evidence to support its use and because of potential side effects [21]. (See "Chronic intestinal pseudo-obstruction: Etiology, clinical manifestations, and diagnosis", section on 'Etiology'.)

Anticholinesterases — We reserve the use of anticholinesterases to patients with acute exacerbation of CIPO who fail to respond to erythromycin. Case reports have suggested that acute exacerbation of intestinal pseudo-obstruction may respond to treatment with neostigmine (0.5 mg intramuscular or intravenous over five minutes with cardiac monitoring)

and its use relieved symptoms and was safe [5,6]. Neostigmine significantly increased antral and intestinal motor activity in patients with diverse upper gastrointestinal dysmotilities, including five patients with intestinal neuropathies or CIPO [22]. (See "Acute colonic pseudo-obstruction (Ogilvie's syndrome)", section on 'Neostigmine'.)

Metoclopramide — Intravenous metoclopramide (10 mg up to four times daily) is an alternative in patients with an acute exacerbation of CIPO who cannot tolerate or fail to respond to both erythromycin and neostigmine. Side effects associated with metoclopramide include central side effects of anxiety, restlessness, and depression, hyperprolactinemia, QT interval prolongation and extrapyramidal side effects, including dystonia and tardive dyskinesia, which have led to a black box warning. A possible history of an extrapyramidal reaction to this agent should be elicited prior to beginning therapy. (See "Treatment of gastroparesis", section on 'Metoclopramide'.)

MANAGEMENT OF INTRACTABLE SYMPTOMS

Decompression and feeding — Enteral nutrition is typically used for neuropathic disorders or in patients in whom the motility disorder is localized to the stomach and duodenum. Percutaneous endoscopic gastro-jejunostomy (PEG-J) decompression can provide an effective means for decompression and enteral nutrition [23]. Alternatively, access to the stomach or small bowel for venting (decompression to relieve symptoms) and feeding may be performed laparoscopically. Enteral feeding may thus facilitate avoidance of total parenteral nutrition-related complications, but is rarely tolerated by patients [24]. (See "Enteral feeding: Gastric versus post-pyloric", section on 'Post-pyloric feeding'.)

Colonoscopic insertion of a gastrostomy tube into the colon to relieve obstructive symptoms has been described in case reports. While the safety and long-term efficacy of this approach remain to be established, improvement in symptoms for up to two years has been reported [25].

Parenteral nutrition — Parenteral nutrition is necessary for patients with severe dysmotility (usually myopathic pseudo-obstruction) and in those who cannot tolerate enteral nutrition. In a large multicenter experience of home parenteral nutrition (HPN) for pediatric patients, the main complications were catheter-related bloodstream infections (1.7/1000 days of parenteral nutrition) and intestinal failure-associated liver disease (20 percent of cohort). Patients in the cohort with CIPO were not at greater risk than other indications [26]. However, patients with CIPO on total parenteral nutrition appear to have lower final heights and body weight Z-scores,

and there may be micronutrient deficiencies transiently while receiving parenteral nutrition [27]. (See "Nutrition support in critically ill patients: An overview".)

Intestinal transplantation — Intestinal transplantation is indicated in patients in whom long-term parenteral nutrition cannot be initiated or continued safely due to complications or with a poor quality-of-life and worsening pain despite aggressive medical management and optimal parenteral nutrition [28-30].

Limited experience suggests that adults with CIPO and irreversible total parenteral nutrition complications benefit from isolated intestinal transplant (associated with different surgical techniques to empty the native stomach) [31].

Long-term survival is achievable with better quality of life and low risk of disease recurrence. Graft rejection, graft-versus-host disease, and immunosuppression-related lymphoproliferative disorders are more common with small intestinal transplantation than after other organ transplants. However, the results of intestinal transplantation have improved over the past decade.

In a prospective study of 55 patients with CIPO (23 children and 32 adults) who underwent intestinal transplantation (43 without a concomitant liver transplant), patient survival at one and five years was 89 percent and 69 percent, respectively, with graft survival of 87 and 56 percent, respectively [32]. Retransplantation was successful in 86 percent. Initially restored nutritional autonomy was sustainable in 23 (70 percent) of 33 long-term survivors with improved quality of life. The remaining 10 recipients required reinstitution of HPN due to allograft enterectomy or gut dysfunction. Overall, this report concluded that long-term survival is achievable after intestinal transplantation, with better quality of life and low risk of disease recurrence. (See "Overview of intestinal and multivisceral transplantation", section on 'Postoperative complications'.)

Although autologous stem cell transplantation and liver transplantation have been used in patients with mitochondrial cytopathy, it is unclear if the benefits outweigh the risks [33,34].

Autologously derived enteric neural stem cells (ENSC) are a possible treatment option for enteric neuropathies in the future. ENSC have been shown to functionally integrate and rescue function in animal models of intestinal disease. However, further studies are required to fully understand the mechanisms of ENSC rescue before this cellular therapy can be applied clinically [35]. (See "Overview of intestinal and multivisceral transplantation".)

Therapies without a clear role

- **Pacing of the intestine and electrical stimulation** – Pacing of the intestine and electrical stimulation of the stomach or intestine are considered investigational at this time, although initial results have been favorable. (See "Electrical stimulation for gastroparesis".)

- **Cannabinoids** – Symptomatic relief has been reported in patients treated with medical cannabis [36]. However, it is important to note that, in general, nonselective cannabinoid receptor agonists generally inhibit gastrointestinal or colonic motility, and there are reports of intestinal intussusception with cannabis use [37].

- **Fecal microbiota transplantation** – A pilot study demonstrated safety of using serial frozen fecal microbiota transplantation, symptom relief (bloating, pain, and reduced intestinal dilatation on CT imaging) in selected patients with CIPO, and improvement in patient tolerance of enteral nutrition delivered via a naso-jejunal tube. Of note, four of the nine patients had previously undergone ileostomy [38]. Additional studies are needed to validate these findings.

- **Resection of localized disease** – In general, non-transplant surgical management of CIPO should be avoided, as it is associated with high postoperative morbidity and mortality rates and frequent re-operation [39]. Resection of localized disease should be avoided in patients with CIPO. Clinical experience suggests that even though the disease may appear to be localized, it usually becomes evident in the remaining bowel, thereby rendering the benefits of a bypass temporary. Repeated surgery also leads to diagnostic confusion, making it difficult to distinguish CIPO from small bowel obstruction. Subtotal enterectomy for pseudo-obstruction has been performed for relief of severe pain associated with markedly distended loops of intestine. In such cases, the patient is committed to total parenteral nutrition for life as a consequence of surgery. Bypass of dilated segments has been suggested for patients with megaduodenum. However, in our experience, this procedure has been ineffective in patients with persistent symptoms.

PROGNOSIS

A number of clinical, histopathologic, and manometric features have been identified as being predictive of outcomes in patients with chronic intestinal pseudo-obstruction (CIPO) (table 1) [40,41]. Long-term outcomes for patients with CIPO are especially poor in children, with 60 to 80 percent requiring parenteral nutrition and a mortality rate ranging from 10 to 40 percent [42-44].

In adults with CIPO, the vast majority of patients have evidence of nutritional compromise, and almost one-third require long-term home parenteral nutrition (HPN) [40]. Mortality rates of approximately 10 percent have been reported in adults, including HPN-related complications in 45 to 80 percent of cases [45]. In one series in adults that included 59 consecutive patients who were followed for a median of 4.6 years, symptoms associated with CIPO became progressively more severe with time [46]. Most patients had undergone a potentially dangerous surgery (mean 2.96 per patient) before a diagnosis was established, underscoring the need to recognize its presentation. Long-term outcomes were poor despite medical and surgical treatment. Approximately one-third of patients required long-term HPN, while about two-thirds had some nutritional limitation. Four patients underwent small bowel transplantation. Overall, 10 percent of patients died of disease-related complications.

SOCIETY GUIDELINE LINKS

Links to society and government-sponsored guidelines from selected countries and regions around the world are provided separately. (See "Society guideline links: Bowel obstruction".)

SUMMARY AND RECOMMENDATIONS

- **Definition** – Chronic intestinal pseudo-obstruction (CIPO) is a syndrome that suggests mechanical bowel obstruction of the small or large bowel in the absence of an anatomic lesion that obstructs the flow of intestinal contents. Segments of affected bowel appear dilated on radiography. (See 'Introduction' above and "Chronic intestinal pseudo-obstruction: Etiology, clinical manifestations, and diagnosis".)

- **Initial management**

  - Initial management of CIPO consists of dietary modification and treatment of the underlying disease.

  - In patients with continued symptoms despite dietary modification or patients with severe symptoms, we suggest prucalopride (**Grade 2C**).

  - In patients with persistent symptoms despite the use of prokinetics, addition of an antiemetic medication may help to relieve symptoms of CIPO. We prefer to use antiemetics such as promethazine and prochlorperazine and reserve the use of 5HT3 antagonists (eg, ondansetron) for patients who cannot tolerate other antiemetic agents, as 5HT3 antagonists can delay colonic transit. (See 'Initial management' above.)

- For patients with CIPO who have steatorrhea, fat-soluble vitamin or vitamin B12 malabsorption, we make a presumptive diagnosis of small intestinal bacterial overgrowth (SIBO) given the limitations of breath tests in patients with severe dysmotility.

  The management of SIBO in patients with CIPO is similar to other patients and consists of antibiotic therapy. Jejunal aspirate cultures are needed if steatorrhea does not respond to empiric antibiotics.

  We avoid the use of octreotide in patients with CIPO since it delays gastric emptying and small bowel transit of solids, which may be deleterious to patients with CIPO. (See 'Additional therapies in selected patients' above and "Small intestinal bacterial overgrowth: Management", section on 'Antibiotic therapy'.)

- **Management of acute exacerbations** – For symptomatic relief in patients with acute exacerbations of CIPO, we suggest intravenous erythromycin rather than other prokinetic agents (**Grade 2C**). We reserve the use of intravenous anticholinesterases (eg, neostigmine) in patients with acute exacerbation of CIPO who fail to respond to erythromycin. (See 'Management of acute exacerbations' above.)

- **Patients with intractable symptoms**

  - Surgery should be performed to provide access to the stomach or small bowel for venting and feeding. Resection of localized disease should be avoided in patients with CIPO.

  - Parenteral nutrition is necessary for patients with severe dysmotility (usually myopathic pseudo-obstruction) and in those who cannot tolerate enteral nutrition.

  - Intestinal transplantation is reserved for patients in whom long-term parenteral nutrition cannot be initiated or continued safely. (See 'Management of intractable symptoms' above.)

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GRAPHICS

Prognostic factors in chronic intestinal pseudo-obstruction

Poor outcome	Good outcome
▪ Myopathy ▪ Early/acute onset ▪ Urinary involvement ▪ Malrotation ▪ Short bowel syndrome ▪ Surgery ▪ Total parenteral nutrition (TPN) ▪ Findings on small intestinal manometry: • Hypomotility • Absence of MMCs • Intestinal phasic and tonic pressure "bursts" • Inadequate intestinal response to a meal (fed pattern)	▪ Sex (male) ▪ No vagal dysfunction or sympathetic dysfunction ▪ Clinical response to cisapride ▪ Normal bowel diameter ▪ Findings on small intestinal manometry: • Presence of MMCs • Motor response to octreotide

MMCs: migrating motor complexes.

Data from: Stanghellini V, Cogliandro RF, De Giorgio R, et al. Natural history of intestinal failure induced by chronic idiopathic intestinal pseudo-obstruction. Transplant Proc 2010; 42:15.

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Contributor Disclosures

Michael Camilleri, MD Equity Ownership/Stock Options: Bilayer Therapeutics [Bile acids in constipation]; Dignify Therapeutics [Spinal cord injury]; Enterin [Constipation in Parkinson disease]; Thelium Therapeutics [Epithelial barrier function]. Patent Holder: 13C-mannitol for permeability measurement [Intestinal diseases]; Capsule for colonic transit by scintigraphy [Colonic motility disorders, including constipation and diarrhea]; Obesity-metabolomics to identify different phenotypes [Obesity subgroups]. Grant/Research/Clinical Trial Support: Allergan [Bile acid malabsorption]; NGM Biopharmaceuticals [Bile acid diarrhea; bile acid malabsorption]; Vanda [Gastroparesis]. Consultant/Advisory Boards: Aclipse Therapeutics [Gastroparesis]; Aditum Bio [Gastroparesis, IBS, IBD]; AEON Pharma [Gastroparesis]; Arena [Visceral pain in GI disorders]; BioKier [Obesity, diabetes]; Coloplast [Colonic motility disorders]; Colospan [Device to measure colon pressure]; Cosmo Pharmaceuticals [Bile acid diarrhea]; Fauna Bio [Intestinal mucosal barrier]; GlaxoSmithKline [Chronic idiopathic constipation]; Invea Therapeutics [IBS-D]; InveniAI [GI motility disorders]; Ironwood [IBS, gastroparesis, bile acid diarrhea]; Kallyope [Obesity, GI function and appetite control]; Novome [GI motility diseases and role of bile acids]; Pfizer [Intestinal epithelial biology and "leaky gut"]; Protagonist Therapeutics [IBS-diarrhea]; QED Therapeutics [Diarrhea]; Sunovion Pharmaceuticals [Gastric functions, satiation, incretins and glycemic control]; Takeda [Gastroparesis screening tool]; VIPUN Medical [Gastric monitoring system]; Virios [HSV-1 and pain in GI diseases]; Zealand Biopharma [Gastroparesis, IBS-D]. All of the relevant financial relationships listed have been mitigated. **Lawrence S Friedman, MD** Other Financial Interest: Elsevier [Gastroenterology]; McGraw-Hill [Gastroenterology]; Wiley [Gastroenterology]. All of the relevant financial relationships listed have been mitigated. **Shilpa Grover, MD, MPH, AGAF** No relevant financial relationship(s) with ineligible companies to disclose.

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