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ABOUT COVER

Peer-reviewer of *World Journal of Clinical Cases*, Dr. Mohamad Adam Bujang is a Research Officer at the Institute for Clinical Research, Ministry of Health, Malaysia. After receiving his Bachelor's degree in Statistics from MARA University of Technology in 2004, Dr. Adam undertook his postgraduate study at the same university, receiving his Master's degree (MBA) in 2008 and his PhD in Information Technology and Quantitative Sciences in 2017. Currently, he works as a biostatistician and researcher in the Clinical Research Centre, Sarawak General Hospital. His ongoing research interests involve such research methodologies as sampling techniques, sample size planning, and statistical analyses. Since 2016, he has served as an active member of the Malaysia Institute of Statistics and the Association of Clinical Registries Malaysia. (L-Editor: Filipodia)

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Latest developments in chronic intestinal pseudo-obstruction

Chang-Zhen Zhu, Hong-Wei Zhao, Hong-Wei Lin, Feng Wang, Yuan-Xin Li

ORCID number: Chang-Zhen Zhu 0000-0003-3712-793X; Hong-Wei Zhao 0000-0003-1130-4264; Hong-Wei Lin 0000-0002-6327-7512; Feng Wang 0000-0003-1385-2694; Yuan-Xin Li 0000-0001-8176-822X.

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Chang-Zhen Zhu, Hong-Wei Zhao, Hong-Wei Lin, Feng Wang, Yuan-Xin Li, Department of Gastrointestinal Surgery, Beijing Tsinghua Changgung Hospital, School of Clinical Medicine, Tsinghua University, Beijing 102218, China

Corresponding author: Yuan-Xin Li, MD, Chief Doctor, Professor, Surgeon, Department of Gastrointestinal, Beijing Tsinghua Changgung Hospital, School of Clinical Medicine, Tsinghua University, No. 168 Litang Road, Changping District, Beijing 102218, China.
liyuanxin1966@163.com

Abstract

Chronic intestinal pseudo-obstruction (CIPO) is a type of intestinal dysfunction presenting as symptoms of intestinal obstruction but without actual mechanical obstruction. An extremely low incidence, non-specific clinical symptoms, strong heterogeneity, and no definitive cause in some patients make CIPO very difficult to diagnose correctly. Imaging and gastrointestinal manometry are commonly used. Most patients have progressive worsening of their symptoms and require intervention, and nutritional assessment and treatment are very important to determine the prognosis. With improvements in surgical techniques, small bowel transplantation is a feasible treatment option for patients with advanced CIPO; however, the long-term prognosis for CIPO patients remains unsatisfactory. Generally, the disease is rare and difficult to diagnose, which leads to clinicians' lack of understanding of the disease and results in a high rate of misdiagnosis. This review describes the characteristics of CIPO and the latest developments in diagnosis and treatment, in detail. The goal of our review is to improve clinicians' understanding of CIPO so that the disease is identified quickly and accurately, and treated as early as possible to improve patients' quality of life.

Key Words: Chronic intestinal pseudo-obstruction; Intestinal obstruction; Enteral nutrition; Parenteral nutrition; Intestinal transplantation

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Core Tip: Chronic intestinal pseudo-obstruction (CIPO) is an intestinal motility disorder caused by neuropathies, myopathies, and mesenchymopathies. CIPO is very difficult to diagnose correctly, leading to most patients experiencing several years from symptom onset to diagnosis. The high misdiagnosis rate relates not only to the characteristics of CIPO itself, but also to the lack of clinicians' understanding of the disease. Using

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published studies, we systematically summarized the diagnosis, treatment, and other information related to CIPO to help clinicians recognize this disease early and minimize patients' suffering.

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INTRODUCTION

Intestinal pseudo-obstruction, first reported by Dudley in 1950^[1], refers to a rare disordered peristalsis characterized by symptoms of intestinal obstruction, but without frank mechanical obstruction^[2]. The disorder is caused by abnormalities of the enteric neuromusculature and/or its autonomic innervation and represents the most severe form of gastrointestinal dysmotility with debilitating and potentially lethal consequences. Chronic intestinal pseudo-obstruction (CIPO) was proposed by Christensen in 1978 to describe intestinal pseudo-obstruction persisting for more than 6 mo^[3]. Due to its rarity and non-specific symptoms, the misdiagnosis rate is extremely high, and CIPO is at an advanced and severe level, when the correct diagnosis is established^[4,5]. We reviewed the most recent literature on CIPO, and comprehensively summarized the etiology, diagnosis, treatment, and prognosis to improve clinicians' understanding of the disease.

EPIDEMIOLOGY

Clinically, CIPO is quite rare, and definitive knowledge regarding its prevalence or incidence is lacking in most countries. According to Vargas, the incidence of pediatric CIPO is estimated at approximately 1 per 40000 live births^[6]. According to data from the American Pseudo-Obstruction and Hirschsprung's Disease Society, an estimated 100 infants are born with congenital pseudo-obstruction every year in the United States^[7]. In 2014, a Japanese survey found that the estimated prevalence was 3.7/1000000, and 56.5% of cases had a neonatal onset; 41 (91.1%) had no pathological abnormalities and were considered idiopathic^[8]. In another survey of 378 institutions belonging to the Japanese Society of Gastroenterology, CIPO prevalence in adult patients was estimated at 1.0 and 0.8 cases per 100000 men and women, respectively. The incidence in the same population was 0.21 and 0.24 cases per 100000 males and females, respectively^[9].

CLASSIFICATION

CIPO is generally divided into congenital, acquired, and idiopathic (no definitive cause) forms^[10]. Most CIPO cases occur in children, are sporadic, and patients have no clear family history. However, some cases with obvious genetic characteristics suggest that CIPO may involve autosomal dominant-, autosomal recessive-, and sex chromosome-related inheritance. CIPO is a common clinical manifestation in many types of mitochondrial myopathies, such as mitochondrial neurogastrointestinal encephalomyopathy (MNGIE), which is caused by a mutation in the *TYMP* gene^[11,12]. Megacystis-microcolon-intestinal hypoperistalsis syndrome (MMIHS) caused by a mutation in the *ACTG2* gene^[13] and chronic atrial and intestinal dysrhythmia syndrome (CAID) caused by mutations in the *SGOL1* gene^[14] also belong to this category. CIPO has also been reported in association with Alpers' disease, which is caused by *POLG* gene mutations or mitochondrial encephalopathy, lactic acidosis, and stroke-like episodes syndrome^[10]. Table 1 shows the details of the congenital forms of CIPO. Patients with hereditary degenerative smooth muscle and enteric nervous system diseases, *i.e.*, familial visceral myopathy and neuropathy, also have CIPO manifestations, but the underlying genes have not been identified.

Table 1 Congenital forms

Genetic mode (Chromosome)	Mutant gene/disease
Autosomal dominant inheritance	SOX10/Waardenburg-Shah syndrome ^[60]
Autosomal recessive inheritance	ACTG2/Megacystis-microcolon-intestinal hypoperistalsis syndrome
	SGOL1/Chronic atrial and intestinal dysrhythmia syndrome
	POLG/Alpers' disease ^[116]
	TYMP, POLG/Mitochondrial neurogastrointestinal encephalomyopathy
	8q23-q24: A new chromosomal localization related to CIPO ^[117]
X-linked recessive	Xq28: <i>Filamin A</i> and <i>L1CAM</i> genes ^[118]

CIPO: Chronic intestinal pseudo-obstruction.

Acquired CIPO is more common in adults and is often secondary to a wide array of diseases, such as systemic neurological, endocrine, and connective tissue diseases, or malignancy. These diseases affect intestinal motility through one or more of the following: autonomic nervous system (stroke, encephalitis, calcification of basal ganglia, orthostatic hypotension, diabetes); intestinal wall nervous system (paraneoplastic syndromes, viral infections, iatrogenic (anthraquinones), diabetes, Hirschsprung's disease, Chagas' disease, Von Recklinghausen's disease); intestinal wall muscle layer (myotonic dystrophy, progressive systemic sclerosis); mixed enteric nervous system and smooth muscle layer (scleroderma, dermatomyositis, amyloidosis, Ehlers-Danlos syndrome, jejunal diverticulosis, radiation enteritis); and unknown mechanisms (hypothyroidism, hypoparathyroidism, pheochromocytoma, clonidine, phenothiazines, antidepressants, antiparkinsonian drugs, antineoplastics, bronchodilators). Neurological diseases such as Parkinson's disease and metabolic diseases such as diabetes affect intestinal function through parasympathetic and/or sympathetic nerves. Paraneoplastic syndromes may cause inflammatory/immune infiltrates of neurons located in submucosal and myenteric ganglia of the enteric nervous system by cellular infiltrates and circulating antineuronal antibodies, resulting in intestinal motility disorders. Myotonic dystrophy or progressive systemic sclerosis mainly damage the enteric smooth muscle cells^[7,15,16]. Autoimmune disorders, collagenopathies, jejunal diverticulosis, and radiation enteritis can alter enteric nerves and smooth muscle cells and even the interstitial cells of Cajal, resulting in a combined neuro-myopathy. Other diseases such as hypothyroidism, hypoparathyroidism, and pheochromocytoma are known causes of CIPO, but the pathogenesis remains unknown. Given that healthy controls rarely harbor viral DNA in the myenteric plexus compared with CIPO patients, many different neurotropic viruses^[17] such as herpesviruses^[18], polyomaviruses (JC virus)^[19], cytomegalovirus^[20,21], and Epstein-Barr virus^[22] might play an etiological role in the development of CIPO.

PATHOLOGY

Congenital and acquired CIPO both constitute three histological categories: neuropathies, myopathies, and mesenchymopathies. More than one pathological type can occur in the same patient simultaneously. (1) Intestinal neuropathies involve neurodegeneration and dysplasia of the enteric nervous system. Neurodegenerative disorders, such as neuronal intranuclear inclusion disease^[23], are characterized by progressive degeneration and disappearance of ganglion cells, leading to a gradual decrease in the number of nerves. In addition to symptoms related directly to CIPO, symptoms also include neurological abnormalities such as ataxia. Dysplastic disorders of the enteric nervous system constitute mainly aganglionosis in Hirschsprung's disease^[24], diffuse intestinal ganglioneuromatosis^[25], intestinal hypoganglionosis^[25], and neurogenic intestinal dysplasia^[26,27], which involve abnormalities in intestinal nerve distribution, development, and quantity. CIPO can also be seen in other diseases, such as intestinal ganglionitis; (2) Intestinal myopathies involve the supernumerary intestinal muscle coat and diffuse abnormalities in muscle layering^[28,29]. Portions of the muscularis propria retain the normal bilayered architecture, but malformed regions contain broad fascicles of smooth muscle that course obliquely or perpendicularly. In

degenerative leiomyopathy, smooth muscle degenerates, is lost, and is then gradually replaced by fibrous connective tissue; CIPO symptoms appear gradually. Other adverse stimuli, such as inflammation of the intestinal wall^[24,30], ischemia, drugs, and immune dysfunction^[4], also cause CIPO in intestinal myopathies; and (3) Mesenchymopathies mainly constitute abnormalities of the interstitial cells of Cajal^[31] and tendinous collagenous tissues of the muscularis propria^[32].

It must be clarified that Hirschsprung's disease is caused by dysplastic disorders of the enteric nervous system, and belongs to the category of intestinal neuropathies, which constitute secondary diseases in CIPO.

SYMPTOMS

Approximately 20% of CIPO cases have clinical manifestations before birth, while 50%–70% of patients show symptoms in the first month after birth. Eighty percent of patients have clinical manifestations at the age of 1 year, and the remaining 20% have sporadic onset before 20 years old^[6,8,33–35]. CIPO may involve any segment of the gastrointestinal tract, and clinical manifestations are mainly related to the location and extent of the involved gastrointestinal tract. Approximately 70% of CIPO patients have esophageal motility disorders^[46]. Generally, the most common symptoms reported by patients are abdominal pain and distension (80%) that is non-colicky, persistent, and aggravated by eating. These symptoms are generally located in the umbilical or upper abdominal regions and gradually spread to involve the whole abdomen. Other symptoms are nausea (75%), vomiting (40%–50%), constipation (40%), and diarrhea (20%–30%)^[5,37–39]. Sub-occlusive episodes can strike in apparently healthy people, but the onset of CIPO is generally insidious, with gastrointestinal symptoms preceding the first acute episode and symptoms worsening significantly during acute episodes, which can last only a few hours^[5,37]. Between acute attacks, patients are asymptomatic or affected by persistent gastrointestinal obstruction^[5,33]. The frequency and severity of acute and intermittent exacerbations are unpredictable, have no detectable cause, and vary widely from patient to patient^[5]. Small intestinal bacterial overgrowth (SIBO) may occur in approximately 30% of patients^[5], which can cause mucosal damage, steatorrhea, diarrhea, and intestinal damage, with chronically dilated bowel loops contributing to malabsorption and vitamin deficiency^[40], which then lead to weight loss. Furthermore, intractable constipation may occur when SIBO is treated with antibiotics. The urinary system is the most common organ involved outside the digestive tract, manifesting mainly as bladder and ureteral dilation associated with a microcolon, a phenotype referred to as MMIHS^[41], which is considered the most severe form of CIPO. Onset is antenatal^[42] and is detected prenatally by ultrasonography in up to 59% of CIPO cases^[43,44]. Patients with CIPO may develop depression^[45] or other psychological disorders due to the long course of the disease and unsatisfactory treatment. Secondary CIPO is common in older people, presenting as the characteristics of the primary disease(s). For example, MNGIE patients not only have intestinal obstruction, but also progressive external ophthalmoplegia, ptosis, and peripheral polyneuropathy^[46].

Due to the non-specific nature of CIPO, even if the identification of CIPO can be improved by clinical features such as extra-intestinal manifestations and malnutrition^[20,47–50], it is still easy to confuse CIPO with other diseases with similar symptoms, such as gastroparesis, functional constipation, cyclic vomiting syndrome, drug toxicity, and hypothyroidism. The reasons are as follows^[51]: (1) Diagnosis still relies on clinical experience rather than on biomarkers of disease; (2) The clinical presentation (intestinal sub-occlusive crisis mimicking a mechanical sub-occlusion) and clinical complexity (*i.e.*, the presence of comorbidities, such as urinary bladder abnormalities or syndromic forms) are factors; and (3) The wide heterogeneity of mechanisms leading to CIPO and related clinical manifestations are additional factors. The diagnosis of CIPO can only be made after several abdominal explorations to exclude mechanical obstruction. The median time for the final correct diagnosis is 8 years, and 88% of patients undergo an average of three non-therapeutic operations^[5].

DIAGNOSIS

Computed tomography (CT) and plain upright abdominal radiography show non-specific obstruction of the digestive tract. Mechanical obstruction can be eliminated by contrast medium examination, but the risk of retaining contrast medium and the

formation of fecal stones, which aggravate obstruction, are concerns. Currently, contrast-enhanced examination has been gradually replaced by high-resolution CT or magnetic resonance imaging (MRI)^[52,53]. CT demonstrates mechanical obstruction, but also provides more information, such as the presence of intestinal adhesions. As a means of assessing and monitoring gastrointestinal motility, cine-MRI can detect subtle contractile impairment of the gut in patients with CIPO, and this method is widely used clinically because of its non-invasive, non-radiation advantages^[54].

Gastrointestinal manometry can effectively evaluate the intensity and coordination of digestive tract contraction^[55,56], which is significant for diagnosis. Abnormal esophageal pressure occurs in approximately 50% of patients, and measuring gastric antral and duodenal pressure is helpful to evaluate motility in the gastric antrum and proximal small intestine, and colonic pressure can directly reflect colonic motility. Some studies have shown that there is no correlation between the results of gastrointestinal manometry and the pathological types of CIPO^[10] and no apparent relationship between the manometric pattern(s) and the type or severity of a patient's symptoms^[57]. However, other studies suggest that antroduodenal manometry may be helpful both in diagnosing CIPO and in indicating the etiology of the condition by showing manometric features suggesting intestinal myogenic or neuropathic processes^[58]. Small intestinal manometry is useful not only as an aid in providing pathophysiologically relevant information regarding the mechanisms underlying dysmotility in CIPO patients (*e.g.*, neuropathic *vs* myopathic patterns), but also in predicting outcomes^[56].

Endoscopy can confirm mechanical obstruction caused by space-occupying lesions and stenosis in the digestive tract. Colonoscopy can also play a role in gastrointestinal decompression. Although capsule endoscopy can provide more information on the digestive tract, this method has potential risks, especially without excluding mechanical obstruction^[59]. The diagnostic value of capsule endoscopy for CIPO is still unclear, and the technique is not currently recommended.

Genetic testing is useful for the diagnosis of special types of diseases associated with CIPO. These diseases include SOX10/Waardenburg-Shah syndrome^[60], *ACTG2*/MMIHS^[13], *SGOL1*/CAID^[14], *POLG*/Alpers' disease, *TYMP* mutation, and *POLG*/MNGIE^[11,12]. When we encounter these diseases, genetic testing can help make the correct diagnosis quickly.

Due to limitations in our understanding of the nerve and muscular layers of the intestinal wall, it is very difficult to interpret the histopathology of CIPO^[61]. It is not uncommon to find no recognizable abnormalities despite a thorough microscopic evaluation of full-thickness bowel biopsies, in children with CIPO. Therefore, full-thickness biopsies of the gastric antrum, small intestine, and colon are of limited value in the diagnosis of CIPO, and this method is not recommended as a routine examination.

Establishing a diagnosis of CIPO is mainly based on clinical manifestations, as well as the presence of extensive intestinal dilatation and multiple gas-liquid levels as imaging findings, and abnormal intestinal manometry, after excluding mechanical obstruction caused by occupying lesions detected by imaging or endoscopy. Diagnostic examination should also identify the pathogenesis of CIPO as clearly as possible, such as with full-thickness biopsies.

TREATMENT

Except for a small number (11%) of asymptomatic patients, most patients have progressive worsening of their symptoms and require medical intervention. Increasing gastrointestinal motility, improving nutritional status, and maintaining the stability of the internal environment are the goals of CIPO therapy^[62]. For patients with secondary CIPO, the primary disease should be actively treated. It is important to be aware that currently, the overall effects of CIPO treatment are unsatisfactory, and the disease course is long; therefore, patients are prone to depression and other psychological disorders^[45,63].

Nutritional support

Please refer to the subsection, "nutritional assessment and support", for details.

Treatment of acute intestinal obstruction

It is crucial to evaluate and maintain the stability of the internal environment. As much as possible, surgical intervention should be minimized because operation can inhibit

intestinal peristalsis, and even induce intestinal failure, which can significantly increase the reoperation rate^[64,65]. However, CIPO patients with Hirschsprung's disease are at risk for colonic volvulus, and timely operation is recommended in this situation^[66].

Analgesic therapy

Visceral pain, especially chronic pain, is a primary concern. Due to their inhibition of gastrointestinal peristalsis, opioids are not recommended. Pain can be treated with low-dose tricyclic antidepressants and gabapentin^[67]; however, as the disease progresses, approximately 25% of patients gradually increase their tolerance to analgesic medications^[5]. It is suggested that surgeons participate in CIPO pain management with pain specialists and psychologists^[10].

Gastrointestinal motility therapy

Currently, there is no evidence to support gastrointestinal motility drugs^[41,68]. The most commonly used drug for promoting gastric motility is erythromycin (1.5-2 g qd for adults; 3-5 mg/kg qd for children)^[5,69,70]. Drugs promoting intestinal peristalsis are octreotide (100 mcg/day, subcutaneous injection)^[71,72] and amoxicillin-clavulanic acid^[73]. There is still insufficient evidence for the treatment of CIPO with metoclopramide and domperidone, and the former carries the risk of tardive dyskinesia when used long-term^[74]. Acetylcholinesterase inhibitors (neostigmine, 8 mg/d, intravenous injection^[75]; pyridostigmine, 20 mg/d, oral administration^[76]) have also been used in adults with CIPO. Prucalopride, a highly selective 5-hydroxytryptamine-4-receptor agonist lacking cardiotoxicity, exerts significant enterokinetic effects^[77]. Cisapride and tegaserod are effective, but these drugs have been banned because of related fatal arrhythmias^[78,79].

Gastrointestinal decompression

Enemas (polyethylene glycol, glycerin), nasogastric tubes, small intestinal tubes, digestive endoscopy, and even surgical fistulation are all optional methods. Colonoscopic decompression has been successfully used in the preoperative treatment of pregnant women with CIPO^[80]. Percutaneous endoscopic gastrojejunostomy fully reduces pressure and relieves abdominal pain, and allows for good nutritional management in patients with CIPO^[81]. Currently, there is no consensus on the timing of decompression by enterostomies in patients with CIPO^[82].

Anti-infection

SIBO is the most common complication of chronic bowel dilatation, and bacterial concentrations are usually higher than 10^3 - 10^5 CFU/mL^[83]. Oral antibiotic therapy is effective, and there are mature antibiotic programs to choose from^[84], such as amoxicillin-clavulanic acid (500 mg, tid), ciprofloxacin (500 mg, bid), doxycycline (100 mg, bid), metronidazole (250 mg, tid), neomycin (500 mg, bid), rifaximin (550 mg, bid), and tetracycline (250 mg, qid)^[85,86]. Antibiotics should be prescribed for 7-10 d each month, with the type of antibiotic changed each month for 5-6 mo^[87].

Fecal bacterial transplantation

Some studies^[88] have shown that fecal bacterial transplantation significantly alleviates patients' abdominal distention and abdominal pain, increases their tolerance for enteral nutrition, and prevents and treats SIBO, indicating that fecal bacterial transplantation may be a new direction in CIPO treatment.

Immunosuppressive therapy

For CIPO patients with significant inflammation/immune reaction in the myenteric ganglion or neuromuscular tract, immunosuppressive agents may reverse the histopathological signs^[89-91]. However, due to a lack of evidence-based medical data, this method should be used with caution.

Allogeneic hematopoietic stem cell transplantation

This therapy definitely improves clinical symptoms in patients with mitochondrial neurogastrointestinal encephalomyopathy, long-term, but the effect in patients with CIPO with intestinal failure is unsatisfactory^[92].

Small intestine/multi-visceral transplantation

Currently, small bowel transplantation secondary to CIPO accounts for approximately

9% of all small bowel transplantations^[93]. The 1-, 3-, and 5-year survival rates between small bowel transplantation secondary to intestinal dysfunction and total small bowel transplantation (75% *vs* 73%, 62% *vs* 61%, and 57% *vs* 55%, respectively) are comparable. Additionally, the incidences of infection and opportunistic complications are similar in CIPO and non-CIPO small bowel transplantation^[94]. Nakamura *et al*^[95] found that the overall 3-year survival rate after small bowel transplantation in patients with total intestinal aganglionosis was 66%, and the longest survival time was 12.8 years. Therefore, for CIPO patients with intestinal failure and potentially fatal complications of parenteral nutrition, small bowel transplantation is a feasible treatment option that can improve long-term prognosis^[93,96].

If other organs are involved, modified multi-visceral transplantation should be performed, such as stomach-duodenum-pancreas-small intestine transplantation, or combined liver and intestine transplantation^[97]. A study of 98 patients undergoing multiple organ transplantation from the University of Miami showed that the 5-year survival rates of patients and grafts were 49% and 47%, respectively^[98]. Considering that, compared with general transplantation, this type of operation is more difficult, more traumatic, and requires more complex postoperative management, this approach is recommended only in experienced transplantation centers.

NUTRITIONAL ASSESSMENT AND SUPPORT

Approximately 2/3 of patients with CIPO have insufficient nutritional intake and may experience significant weight loss^[10]. Therefore, strict and systematic nutritional evaluation and selecting appropriate nutritional supports are essential to improve patients' quality of life and long-term prognosis. However, no evidence-based consensus proposals are available for nutritional management in patients with CIPO^[99].

Nutritional assessment

A strict and systematic nutritional assessment of patients with CIPO should be performed by experienced nutritionists. Macro measurements such as weight, body mass index, and eating habits, and laboratory measurements such as serum albumin, prealbumin, lymphocyte count, and C-reactive protein are recommended. Secondary to insufficient intake and/or absorption obstacles in some patients, the concentrations of serum calcium, iron, vitamin B12, folic acid, fat-soluble vitamins, thiamine, and niacinamide may be abnormal; therefore, levels should be measured and corrected, as needed.

Nutritional support

Dietary support: Nutritionists provide nutritional education and create personalized diet plans for patients. Generally, oral intake is most desirable, and smaller, more frequent meals (5–6 times/day) are recommended^[65,100–103]. The ideal diet is a liquid formulation containing protein and high fat; high fiber diets should be restricted or even avoided. Patients' levels of water-soluble vitamins, minerals, and electrolytes should be regularly measured and supplemented, as needed. If SIBO occurs, it must be remembered that the levels of fat-soluble vitamins and vitamin B12 should be measured^[100–102,104]. If necessary, an elemental diet and dietary supplements rich in medium-chain triglycerides can be used^[61].

Enteral nutrition: Enteral nutrition can be considered if oral feeding cannot meet the patient's nutritional requirements. Standard, non-elemental enteral nutrition can generally meet the nutritional needs of most patients with CIPO. Nasogastric tubes or gastrostomy tubes can be used for continuous and small-dose feeding, if needed^[105]. Nasojejunostomy or jejunostomy tubes can work well if gastroparesis occurs. Compared with one-time high-dose feeding, continuous, low-dose, or periodic feeding is more easily accepted by patients^[106].

Parenteral nutrition: Approximately 60%–80% of patients rely on parenteral nutritional support to different degrees^[97–108], and home parenteral nutrition is used by approximately 20% of patients. Approximately 2/3 of children with CIPO rely on parenteral nutrition for sufficient energy intake^[109]. If patients are exclusively dependent on parenteral nutrition, total energy intake is calculated as approximately 25 kcal/kg/day, and lipids should supply approximately 30% of total parenteral calories with 1.0–1.5 g/kg/day of protein and the remaining calories provided by

glucose^[110,111]. Amiot *et al*^[47] found that parenteral nutrition can effectively maintain patients' body weight and relieve symptoms. The 1-, 5-, 10-, and 15-year survival rates of patients with CIPO supported by parenteral nutrition were 94%, 78%, 75%, and 68%, respectively, and approximately 2/3 of patients used home parenteral nutrition for 15 years. However, 90% of deaths in patients with CIPO were related to parenteral nutrition-related complications, in one study^[55]. Other studies reported that the mortality rate of children with CIPO ranged from 4.7% to 15.29%^[107,112]. Metabolic complications include metabolic bone disease, electrolyte abnormalities, dehydration, and intestinal failure-associated liver disease. Infectious complications include catheter-related blood stream infections, and exit site. Mechanical complications include catheter breaks, catheter occlusions, thrombosis, especially deep vein thrombosis, superior vena cava syndrome, and air embolism^[113]. Therefore, maximizing oral intake in patients with^[62] CIPO is recommended to minimize or avoid the use of parenteral nutrition.

PROGNOSIS

The long-term prognosis of patients with CIPO is unsatisfactory. Intestinal myopathies, esophageal involvement, inadequate response of the digestive tract to food stimulation, deficiencies in the migrating motor complex during fasting, concurrent urinary retention, and concurrent intestinal malrotation are poor prognostic factors^[35,114,115]; 10%–25% of children with CIPO die before adulthood. Parenteral nutrition combined with oral feeding can significantly improve patients' survival rates, and oral feeding is an independent predictor of long-term prognosis^[89].

CONCLUSION

CIPO is a rare disorder of intestinal function characterized by abnormal intestinal peristalsis. The symptoms, signs, and imaging features all indicate mechanical intestinal obstruction, and the misdiagnosis rate is very high. Clinicians should inquire about the history of disease and physically examine patients carefully. Once the possibility of organic obstruction is eliminated, the possibility of CIPO should be considered. Although no studies have evaluated the relationship between early diagnosis and prognosis, a greater awareness of CIPO would indeed help reduce unnecessary surgical procedures, and full thickness biopsies at an early and potentially curable stage of the disease are recommended to evaluate the gut nervous layer, identify pathology, and to provide targeted treatment as quickly as possible.

REFERENCES

- 1 **Dudley HA**, Sinclair IS, McLaren IF, McNair TJ, Newsam JE. Intestinal pseudo-obstruction. *J R Coll Surg Edinb* 1958; **3**: 206-217 [PMID: [13514744](#)]
- 2 **Rudolph CD**, Hyman PE, Altschuler SM, Christensen J, Colletti RB, Cucchiara S, Di Lorenzo C, Flores AF, Hillemeier AC, McCallum RW, Vanderhoof JA. Diagnosis and treatment of chronic intestinal pseudo-obstruction in children: report of consensus workshop. *J Pediatr Gastroenterol Nutr* 1997; **24**: 102-112 [PMID: [9093995](#) DOI: [10.1097/00005176-199701000-00021](#)]
- 3 **Faulk DL**, Anuras S, Christensen J. Chronic intestinal pseudoobstruction. *Gastroenterology* 1978; **74**: 922-931 [PMID: [346432](#)]
- 4 **Connor FL**, Di Lorenzo C. Chronic intestinal pseudo-obstruction: assessment and management. *Gastroenterology* 2006; **130**: S29-S36 [PMID: [16473068](#) DOI: [10.1053/j.gastro.2005.06.081](#)]
- 5 **Stanghellini V**, Cogliandro RF, De Giorgio R, Barbara G, Morselli-Labate AM, Cogliandro L, Corinaldesi R. Natural history of chronic idiopathic intestinal pseudo-obstruction in adults: a single center study. *Clin Gastroenterol Hepatol* 2005; **3**: 449-458 [PMID: [15880314](#) DOI: [10.1016/s1542-3565\(04\)00675-5](#)]
- 6 **Vargas JH**, Sachs P, Ament ME. Chronic intestinal pseudo-obstruction syndrome in pediatrics. Results of a national survey by members of the North American Society of Pediatric Gastroenterology and Nutrition. *J Pediatr Gastroenterol Nutr* 1988; **7**: 323-332 [PMID: [3290417](#)]
- 7 **Di Lorenzo C**. Pseudo-obstruction: current approaches. *Gastroenterology* 1999; **116**: 980-987 [PMID: [10092321](#) DOI: [10.1016/s0016-5085\(99\)70082-x](#)]
- 8 **Muto M**, Matsufuji H, Tomomasa T, Nakajima A, Kawahara H, Ida S, Ushijima K, Kubota A,

- Mushiaki S, Taguchi T. Pediatric chronic intestinal pseudo-obstruction is a rare, serious, and intractable disease: a report of a nationwide survey in Japan. *J Pediatr Surg* 2014; **49**: 1799-1803 [PMID: [25487487](#) DOI: [10.1016/j.jpedsurg.2014.09.025](#)]
- 9 Iida H, Ohkubo H, Inamori M, Nakajima A, Sato H. Epidemiology and clinical experience of chronic intestinal pseudo-obstruction in Japan: a nationwide epidemiologic survey. *J Epidemiol* 2013; **23**: 288-294 [PMID: [23831693](#) DOI: [10.2188/jea.je20120173](#)]
- 10 Ahmed S, Sharman T. Intestinal Pseudo-Obstruction. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing [PMID: [32809504](#)]
- 11 Nishino I, Spinazzola A, Hirano M. Thymidine phosphorylase gene mutations in MNGIE, a human mitochondrial disorder. *Science* 1999; **283**: 689-692 [PMID: [9924029](#) DOI: [10.1126/science.283.5402.689](#)]
- 12 Van Goethem G, Schwartz M, Löfgren A, Dermaut B, Van Broeckhoven C, Vissing J. Novel POLG mutations in progressive external ophthalmoplegia mimicking mitochondrial neurogastrointestinal encephalomyopathy. *Eur J Hum Genet* 2003; **11**: 547-549 [PMID: [12825077](#) DOI: [10.1038/sj.ejhg.5201002](#)]
- 13 Thorson W, Diaz-Horta O, Foster J 2nd, Spiliopoulos M, Quintero R, Farooq A, Blanton S, Tekin M. De novo ACTG2 mutations cause congenital distended bladder, microcolon, and intestinal hypoperistalsis. *Hum Genet* 2014; **133**: 737-742 [PMID: [24337657](#) DOI: [10.1007/s00439-013-1406-0](#)]
- 14 Chetaille P, Preuss C, Burkhard S, Côté JM, Houde C, Castilloux J, Piché J, Gosset N, Leclerc S, Wünnemann F, Thibeault M, Gagnon C, Galli A, Tuck E, Hickson GR, El Amine N, Boufaied I, Lemyre E, de Santa Barbara P, Faure S, Jonzon A, Cameron M, Dietz HC, Gallo-McFarlane E, Benson DW, Moreau C, Labuda D; FORGE Canada Consortium, Zhan SH, Shen Y, Jomphe M, Jones SJ, Bakkers J, Andelfinger G. Mutations in SGOL1 cause a novel cohesinopathy affecting heart and gut rhythm. *Nat Genet* 2014; **46**: 1245-1249 [PMID: [25282101](#) DOI: [10.1038/ng.3113](#)]
- 15 Stanghellini V, Corinaldesi R, Barbara L. Pseudo-obstruction syndromes. *Baillieres Clin Gastroenterol* 1988; **2**: 225-254 [PMID: [3289641](#) DOI: [10.1016/0950-3528\(88\)90029-2](#)]
- 16 Stanghellini V, Cogliandro RF, de Giorgio R, Barbara G, Salvioli B, Corinaldesi R. Chronic intestinal pseudo-obstruction: manifestations, natural history and management. *Neurogastroenterol Motil* 2007; **19**: 440-452 [PMID: [17564625](#) DOI: [10.1111/j.1365-2982.2007.00902.x](#)]
- 17 De Giorgio R, Ricciardiello L, Naponelli V, Selgrad M, Piazzi G, Felicani C, Serra M, Fronzoni L, Antonucci A, Cogliandro RF, Barbara G, Corinaldesi R, Tonini M, Knowles CH, Stanghellini V. Chronic intestinal pseudo-obstruction related to viral infections. *Transplant Proc* 2010; **42**: 9-14 [PMID: [20172270](#) DOI: [10.1016/j.transproceed.2009.12.014](#)]
- 18 Pavone S, Sforna M, Gialletti R, Prato S, Marenzoni ML, Mandara MT. Extensive myenteric ganglionitis in a case of equine chronic intestinal pseudo-obstruction associated with EHV-1 infection. *J Comp Pathol* 2013; **148**: 289-293 [PMID: [22935089](#) DOI: [10.1016/j.jcpa.2012.07.004](#)]
- 19 Sinagra E, Raimondo D, Gallo E, Calvaruso M, Lentini VL, Cannizzaro A, Linea C, Giunta M, Montalbano LM, D'Amico G, Rizzo AG. Could JC virus be linked to chronic idiopathic intestinal pseudo-obstruction? *Clin J Gastroenterol* 2020; **13**: 377-381 [PMID: [31728918](#) DOI: [10.1007/s12328-019-01069-4](#)]
- 20 Sonsino E, Mouy R, Foucaud P, Cezard JP, Aigrain Y, Bocquet L, Navarro J. Intestinal pseudoobstruction related to cytomegalovirus infection of myenteric plexus. *N Engl J Med* 1984; **311**: 196-197 [PMID: [6330552](#) DOI: [10.1056/NEJM198407193110319](#)]
- 21 Atego S, Turck D, Gottrand F, Bonneville M, Wattré P, Lecomte-Houcke M, Farriaux JP. Chronic intestinal pseudo-obstruction associated with cytomegalovirus infection in an infant. *J Pediatr Gastroenterol Nutr* 1996; **23**: 457-460 [PMID: [8956187](#) DOI: [10.1097/00005176-199611000-00018](#)]
- 22 Besnard M, Faure C, Fromont-Hankard G, Ansart-Pirenne H, Peuchmaur M, Cezard JP, Navarro J. Intestinal pseudo-obstruction and acute pandysautonomia associated with Epstein-Barr virus infection. *Am J Gastroenterol* 2000; **95**: 280-284 [PMID: [10638598](#) DOI: [10.1111/j.1572-0241.2000.01709.x](#)]
- 23 Kapur RP. Pathology of Intestinal Motor Disorders in Children. *Surg Pathol Clin* 2010; **3**: 711-741 [PMID: [26839228](#) DOI: [10.1016/j.path.2010.06.005](#)]
- 24 O'Donnell AM, Puri P. Skip segment Hirschsprung's disease: a systematic review. *Pediatr Surg Int* 2010; **26**: 1065-1069 [PMID: [20714729](#) DOI: [10.1007/s00383-010-2692-4](#)]
- 25 Dingemann J, Puri P. Isolated hypoganglionosis: systematic review of a rare intestinal innervation defect. *Pediatr Surg Int* 2010; **26**: 1111-1115 [PMID: [20721562](#) DOI: [10.1007/s00383-010-2693-3](#)]
- 26 Nezelof C, Guy-Grand D, Thomine E. [Megacolon with hyperplasia of the myenteric plexus. An anatomo-clinical entity, apropos of 3 cases]. *Presse Med* 1970; **78**: 1501-1506 [PMID: [5429378](#)]
- 27 Meier-Ruge W. [Casuistic of colon disorder with symptoms of Hirschsprung's disease (author's transl)]. *Verh Dtsch Ges Pathol* 1971; **55**: 506-510 [PMID: [4130757](#)]
- 28 Kapur RP, Correa H. Architectural malformation of the muscularis propria as a cause for intestinal pseudo-obstruction: two cases and a review of the literature. *Pediatr Dev Pathol* 2009; **12**: 156-164

- [PMID: 18788889 DOI: 10.2350/08-07-0495.1]
- 29 **Smith VV**, Milla PJ. Histological phenotypes of enteric smooth muscle disease causing functional intestinal obstruction in childhood. *Histopathology* 1997; **31**: 112-122 [PMID: 9279561 DOI: 10.1046/j.1365-2559.1997.2250839.x]
 - 30 **Oton E**, Moreira V, Redondo C, Lopez-San-Roman A, Foruny JR, Plaza G, de Vicente E, Quijano Y. Chronic intestinal pseudo-obstruction due to lymphocytic leiomyositis: is there a place for immunomodulatory therapy? *Gut* 2005; **54**: 1343-1344 [PMID: 16099803 DOI: 10.1136/gut.2005.071811]
 - 31 **Negreanu LM**, Assor P, Mateescu B, Cirstoiu C. Interstitial cells of Cajal in the gut--a gastroenterologist's point of view. *World J Gastroenterol* 2008; **14**: 6285-6288 [PMID: 19009640 DOI: 10.3748/wjg.14.6285]
 - 32 **Bruhlin-Feichter S**, Meier-Ruge W, Martucciello G, Bruder E. Connective tissue in gut development: a key player in motility and in intestinal desmosis. *Eur J Pediatr Surg* 2012; **22**: 445-459 [PMID: 22903251 DOI: 10.1055/s-0032-1322544]
 - 33 **Heneyke S**, Smith VV, Spitz L, Milla PJ. Chronic intestinal pseudo-obstruction: treatment and long term follow up of 44 patients. *Arch Dis Child* 1999; **81**: 21-27 [PMID: 10373127 DOI: 10.1136/adc.81.1.21]
 - 34 **Mousa H**, Hyman PE, Cocjin J, Flores AF, Di Lorenzo C. Long-term outcome of congenital intestinal pseudoobstruction. *Dig Dis Sci* 2002; **47**: 2298-2305 [PMID: 12395903 DOI: 10.1023/a:1020199614102]
 - 35 **Diamanti A**, Fusaro F, Caldaro T, Capriati T, Candusso M, Nobili V, Borrelli O. Pediatric Intestinal Pseudo-obstruction: Impact of Neonatal and Later Onset on Clinical and Nutritional Outcomes. *J Pediatr Gastroenterol Nutr* 2019; **69**: 212-217 [PMID: 31058770 DOI: 10.1097/MPG.0000000000002373]
 - 36 **Amiot A**, Joly F, Cazals-Hatem D, Merrouche M, Jouet P, Coffin B, Bouhnik Y. Prognostic yield of esophageal manometry in chronic intestinal pseudo-obstruction: a retrospective cohort of 116 adult patients. *Neurogastroenterol Motil* 2012; **24**: 1008-e542 [PMID: 22762287 DOI: 10.1111/j.1365-2982.2012.01973.x]
 - 37 **El-Chammas K**, Sood MR. Chronic Intestinal Pseudo-obstruction. *Clin Colon Rectal Surg* 2018; **31**: 99-107 [PMID: 29487492 DOI: 10.1055/s-0037-1609024]
 - 38 **Zenzeri L**, Tambucci R, Quitadamo P, Giorgio V, De Giorgio R, Di Nardo G. Update on chronic intestinal pseudo-obstruction. *Curr Opin Gastroenterol* 2020; **36**: 230-237 [PMID: 32073506 DOI: 10.1097/MOG.0000000000000630]
 - 39 **Mann SD**, Debinski HS, Kamm MA. Clinical characteristics of chronic idiopathic intestinal pseudo-obstruction in adults. *Gut* 1997; **41**: 675-681 [PMID: 9414977 DOI: 10.1136/gut.41.5.675]
 - 40 **Cucchiara S**, Borrelli O. Nutritional challenge in pseudo-obstruction: the bridge between motility and nutrition. *J Pediatr Gastroenterol Nutr* 2009; **48** Suppl 2: S83-S85 [PMID: 19300134 DOI: 10.1097/MPG.0b013e3181a15bfc]
 - 41 **Yeung AK**, Di Lorenzo C. Primary gastrointestinal motility disorders in childhood. *Minerva Pediatr* 2012; **64**: 567-584 [PMID: 23108319]
 - 42 **Kubota A**, Okuyama H, Takahashi T, Kawahara H, Nakai H, Yoshida H, Takama Y, Nakacyo M, Ida S. A case of CIIPS presenting with bowel obstruction secondary to malrotation as the first symptom. *Jpn J Pediatr Surg* 2005; **37**: 824-831
 - 43 **Glassman M**, Spivak W, Mininberg D, Madara J. Chronic idiopathic intestinal pseudoobstruction: a commonly misdiagnosed disease in infants and children. *Pediatrics* 1989; **83**: 603-608 [PMID: 2928002]
 - 44 **Lapointe SP**, Rivet C, Goulet O, Fékété CN, Lortat-Jacob S. Urological manifestations associated with chronic intestinal pseudo-obstructions in children. *J Urol* 2002; **168**: 1768-1770 [PMID: 12352356 DOI: 10.1097/01.ju.0000028495.91112.98]
 - 45 **Cogliandro RF**, Antonucci A, De Giorgio R, Barbara G, Cremon C, Cogliandro L, Frisoni C, Pezzilli R, Morselli-Labate AM, Corinaldesi R, Stanghellini V. Patient-reported outcomes and gut dysmotility in functional gastrointestinal disorders. *Neurogastroenterol Motil* 2011; **23**: 1084-1091 [PMID: 21917083 DOI: 10.1111/j.1365-2982.2011.01783.x]
 - 46 **Lara MC**, Valentino ML, Torres-Torronteras J, Hirano M, Martí R. Mitochondrial neurogastrointestinal encephalomyopathy (MNGIE): biochemical features and therapeutic approaches. *Biosci Rep* 2007; **27**: 151-163 [PMID: 17549623 DOI: 10.1007/s10540-007-9043-2]
 - 47 **Amiot A**, Joly F, Alves A, Panis Y, Bouhnik Y, Messing B. Long-term outcome of chronic intestinal pseudo-obstruction adult patients requiring home parenteral nutrition. *Am J Gastroenterol* 2009; **104**: 1262-1270 [PMID: 19367271 DOI: 10.1038/ajg.2009.58]
 - 48 **Lindberg G**, Iwarzon M, Tornblom H. Clinical features and long-term survival in chronic intestinal pseudo-obstruction and enteric dysmotility. *Scand J Gastroenterol* 2009; **44**: 692-699 [PMID: 19308797 DOI: 10.1080/00365520902839642]
 - 49 **De Giorgio R**, Stanghellini V, Barbara G, Corinaldesi R, De Ponti F, Tonini M, Bassotti G, Sternini C. Primary enteric neuropathies underlying gastrointestinal motor dysfunction. *Scand J*

- Gastroenterol* 2000; **35**: 114-122 [PMID: [10720107](#) DOI: [10.1080/003655200750024263](#)]
- 50 **De Giorgio R**, Sarnelli G, Corinaldesi R, Stanghellini V. Advances in our understanding of the pathology of chronic intestinal pseudo-obstruction. *Gut* 2004; **53**: 1549-1552 [PMID: [15479666](#) DOI: [10.1136/gut.2004.043968](#)]
- 51 **Pironi L**, Sasdelli AS. Management of the Patient with Chronic Intestinal Pseudo-Obstruction and Intestinal Failure. *Gastroenterol Clin North Am* 2019; **48**: 513-524 [PMID: [31668180](#) DOI: [10.1016/j.gtc.2019.08.005](#)]
- 52 **Merlin A**, Soyer P, Boudiaf M, Hamzi L, Rymer R. Chronic intestinal pseudo-obstruction in adult patients: multidetector row helical CT features. *Eur Radiol* 2008; **18**: 1587-1595 [PMID: [18357454](#) DOI: [10.1007/s00330-008-0913-1](#)]
- 53 **Menys A**, Butt S, Emmanuel A, Plumb AA, Fikree A, Knowles C, Atkinson D, Zarate N, Halligan S, Taylor SA. Comparative quantitative assessment of global small bowel motility using magnetic resonance imaging in chronic intestinal pseudo-obstruction and healthy controls. *Neurogastroenterol Motil* 2016; **28**: 376-383 [PMID: [26661570](#) DOI: [10.1111/nmo.12735](#)]
- 54 **Fuyuki A**, Ohkubo H, Higurashi T, Iida H, Inoh Y, Inamori M, Nakajima A. Clinical importance of cine-MRI assessment of small bowel motility in patients with chronic intestinal pseudo-obstruction: a retrospective study of 33 patients. *J Gastroenterol* 2017; **52**: 577-584 [PMID: [27549243](#) DOI: [10.1007/s00535-016-1251-8](#)]
- 55 **Goulet O**, Jobert-Giraud A, Michel JL, Jaubert F, Lortat-Jacob S, Colomb V, Cuenod-Jabri B, Jan D, Brousse N, Gaillard D, Nihoul-Fékété C, Ricour C. Chronic intestinal pseudo-obstruction syndrome in pediatric patients. *Eur J Pediatr Surg* 1999; **9**: 83-89 [PMID: [10342114](#) DOI: [10.1055/s-2008-1072218](#)]
- 56 **Fell JM**, Smith VV, Milla PJ. Infantile chronic idiopathic intestinal pseudo-obstruction: the role of small intestinal manometry as a diagnostic tool and prognostic indicator. *Gut* 1996; **39**: 306-311 [PMID: [8977348](#) DOI: [10.1136/gut.39.2.306](#)]
- 57 **Stanghellini V**, Camilleri M, Malagelada JR. Chronic idiopathic intestinal pseudo-obstruction: clinical and intestinal manometric findings. *Gut* 1987; **28**: 5-12 [PMID: [3817584](#) DOI: [10.1136/gut.28.1.5](#)]
- 58 **Cucchiara S**, Annese V, Minella R, Franco MT, Iervolino C, Emiliano M, Auricchio S. Antroduodenal manometry in the diagnosis of chronic idiopathic intestinal pseudoobstruction in children. *J Pediatr Gastroenterol Nutr* 1994; **18**: 294-305 [PMID: [8057211](#) DOI: [10.1097/00005176-199404000-00008](#)]
- 59 **Malagelada C**, De Lorio F, Seguí S, Mendez S, Drozdal M, Vitria J, Radeva P, Santos J, Accarino A, Malagelada JR, Azpiroz F. Functional gut disorders or disordered gut function? Small bowel dysmotility evidenced by an original technique. *Neurogastroenterol Motil* 2012; **24**: 223-228, e104-e105 [PMID: [22129212](#) DOI: [10.1111/j.1365-2982.2011.01823.x](#)]
- 60 **Pingault V**, Girard M, Bondurand N, Dorkins H, Van Maldergem L, Mowat D, Shimotake T, Verma I, Baumann C, Goossens M. SOX10 mutations in chronic intestinal pseudo-obstruction suggest a complex physiopathological mechanism. *Hum Genet* 2002; **111**: 198-206 [PMID: [12189494](#) DOI: [10.1007/s00439-002-0765-8](#)]
- 61 **Knowles CH**, De Giorgio R, Kapur RP, Bruder E, Farrugia G, Geboes K, Lindberg G, Martin JE, Meier-Ruge WA, Milla PJ, Smith VV, Vandervinden JM, Veress B, Wedel T. The London Classification of gastrointestinal neuromuscular pathology: report on behalf of the Gastro 2009 International Working Group. *Gut* 2010; **59**: 882-887 [PMID: [20581236](#) DOI: [10.1136/gut.2009.200444](#)]
- 62 **Joly F**, Amiot A, Messing B. Nutritional support in the severely compromised motility patient: when and how? *Gastroenterol Clin North Am* 2011; **40**: 845-851 [PMID: [22100122](#) DOI: [10.1016/j.gtc.2011.09.010](#)]
- 63 **Lyford G**, Foxx-Orenstein A. Chronic Intestinal Pseudoobstruction. *Curr Treat Options Gastroenterol* 2004; **7**: 317-325 [PMID: [15238207](#) DOI: [10.1007/s11938-004-0018-0](#)]
- 64 **Sabbagh C**, Amiot A, Maggiori L, Corcos O, Joly F, Panis Y. Non-transplantation surgical approach for chronic intestinal pseudo-obstruction: analysis of 63 adult consecutive cases. *Neurogastroenterol Motil* 2013; **25**: e680-e686 [PMID: [23895212](#) DOI: [10.1111/nmo.12191](#)]
- 65 **Murr MM**, Sarr MG, Camilleri M. The surgeon's role in the treatment of chronic intestinal pseudoobstruction. *Am J Gastroenterol* 1995; **90**: 2147-2151 [PMID: [8540505](#)]
- 66 **Altaf MA**, Werlin SL, Sato TT, Rudolph CD, Sood MR. Colonic volvulus in children with intestinal motility disorders. *J Pediatr Gastroenterol Nutr* 2009; **49**: 59-62 [PMID: [19465873](#) DOI: [10.1097/MPG.0b013e3181879eb5](#)]
- 67 **Rosner H**, Rubin L, Kestenbaum A. Gabapentin adjunctive therapy in neuropathic pain states. *Clin J Pain* 1996; **12**: 56-58 [PMID: [8722736](#) DOI: [10.1097/00002508-199603000-00010](#)]
- 68 **Di Nardo G**, Di Lorenzo C, Lauro A, Stanghellini V, Thapar N, Karunaratne TB, Volta U, De Giorgio R. Chronic intestinal pseudo-obstruction in children and adults: diagnosis and therapeutic options. *Neurogastroenterol Motil* 2017; **29** [PMID: [27683196](#) DOI: [10.1111/nmo.12945](#)]
- 69 **Di Lorenzo C**, Flores AF, Tomomasa T, Hyman PE. Effect of erythromycin on antroduodenal

- motility in children with chronic functional gastrointestinal symptoms. *Dig Dis Sci* 1994; **39**: 1399-1404 [PMID: 8026249 DOI: 10.1007/BF02088040]
- 70 **Emmanuel AV**, Shand AG, Kamm MA. Erythromycin for the treatment of chronic intestinal pseudo-obstruction: description of six cases with a positive response. *Aliment Pharmacol Ther* 2004; **19**: 687-694 [PMID: 15023171 DOI: 10.1111/j.1365-2036.2004.01900.x]
 - 71 **Di Lorenzo C**, Lucanto C, Flores AF, Idries S, Hyman PE. Effect of octreotide on gastrointestinal motility in children with functional gastrointestinal symptoms. *J Pediatr Gastroenterol Nutr* 1998; **27**: 508-512 [PMID: 9822313 DOI: 10.1097/00005176-199811000-00002]
 - 72 **Soudah HC**, Hasler WL, Owyang C. Effect of octreotide on intestinal motility and bacterial overgrowth in scleroderma. *N Engl J Med* 1991; **325**: 1461-1467 [PMID: 1944424 DOI: 10.1056/NEJM199111213252102]
 - 73 **Gomez R**, Fernandez S, Aspirot A, Punati J, Skaggs B, Mousa H, Di Lorenzo C. Effect of amoxicillin/clavulanate on gastrointestinal motility in children. *J Pediatr Gastroenterol Nutr* 2012; **54**: 780-784 [PMID: 22584747 DOI: 10.1097/MPG.0b013e31824204e4]
 - 74 **Rao AS**, Camilleri M. Review article: metoclopramide and tardive dyskinesia. *Aliment Pharmacol Ther* 2010; **31**: 11-19 [PMID: 19886950 DOI: 10.1111/j.1365-2036.2009.04189.x]
 - 75 **Calvet X**, Martinez JM, Martinez M. Repeated neostigmine dosage as palliative treatment for chronic colonic pseudo-obstruction in a patient with autonomic paraneoplastic neuropathy. *Am J Gastroenterol* 2003; **98**: 708-709 [PMID: 12650823 DOI: 10.1111/j.1572-0241.2003.07313.x]
 - 76 **O'Dea CJ**, Brookes JH, Wattchow DA. The efficacy of treatment of patients with severe constipation or recurrent pseudo-obstruction with pyridostigmine. *Colorectal Dis* 2010; **12**: 540-548 [PMID: 19508545 DOI: 10.1111/j.1463-1318.2009.01838.x]
 - 77 **Emmanuel AV**, Kamm MA, Roy AJ, Kerstens R, Vandeplasse L. Randomised clinical trial: the efficacy of prucalopride in patients with chronic intestinal pseudo-obstruction--a double-blind, placebo-controlled, cross-over, multiple n = 1 study. *Aliment Pharmacol Ther* 2012; **35**: 48-55 [PMID: 22061077 DOI: 10.1111/j.1365-2036.2011.04907.x]
 - 78 **Di Lorenzo C**, Reddy SN, Villanueva-Meyer J, Mena I, Martin S, Hyman PE. Cisapride in children with chronic intestinal pseudoobstruction. An acute, double-blind, crossover, placebo-controlled trial. *Gastroenterology* 1991; **101**: 1564-1570 [PMID: 1955122 DOI: 10.1016/0016-5085(91)90393-y]
 - 79 **Raphael BP**, Nurko S, Jiang H, Hart K, Kamin DS, Jaksic T, Duggan C. Cisapride improves enteral tolerance in pediatric short-bowel syndrome with dysmotility. *J Pediatr Gastroenterol Nutr* 2011; **52**: 590-594 [PMID: 21502831 DOI: 10.1097/MPG.0b013e3181fe2d7a]
 - 80 **Kim JS**, Lee BI, Kim BW, Choi H, Lee YS, Maeng L. Repetitive Colonoscopic Decompression as a Bridge Therapy before Surgery in a Pregnant Patient with Chronic Intestinal Pseudo-Obstruction. *Clin Endosc* 2013; **46**: 591-594 [PMID: 24143328 DOI: 10.5946/ce.2013.46.5.591]
 - 81 **Ohkubo H**, Fuyuki A, Arimoto J, Higurashi T, Nonaka T, Inoh Y, Iida H, Inamori M, Kaneda T, Nakajima A. Efficacy of percutaneous endoscopic gastro-jejunostomy (PEG-J) decompression therapy for patients with chronic intestinal pseudo-obstruction (CIPO). *Neurogastroenterol Motil* 2017; **29** [PMID: 28631871 DOI: 10.1111/nmo.13127]
 - 82 **Youssef NN**, Barksdale Jr E, Griffiths JM, Flores AF, Di Lorenzo C. Management of intractable constipation with antegrade enemas in neurologically intact children. *J Pediatr Gastroenterol Nutr* 2002; **34**: 402-405 [PMID: 11930097 DOI: 10.1097/00005176-200204000-00016]
 - 83 **Khoshini R**, Dai SC, Lezcano S, Pimentel M. A systematic review of diagnostic tests for small intestinal bacterial overgrowth. *Dig Dis Sci* 2008; **53**: 1443-1454 [PMID: 17990113 DOI: 10.1007/s10620-007-0065-1]
 - 84 **Singh VV**, Toskes PP. Small bowel bacterial overgrowth: presentation, diagnosis, and treatment. *Curr Gastroenterol Rep* 2003; **5**: 365-372 [PMID: 12959716 DOI: 10.1007/s11894-003-0048-0]
 - 85 **Shah SC**, Day LW, Somsouk M, Sewell JL. Meta-analysis: antibiotic therapy for small intestinal bacterial overgrowth. *Aliment Pharmacol Ther* 2013; **38**: 925-934 [PMID: 24004101 DOI: 10.1111/apt.12479]
 - 86 **Ghoneum M**, Cooper EL, Sadek I. Variability of natural killer cell activity in anuran amphibians. *Dev Comp Immunol* 1990; **14**: 359-365 [PMID: 2210011 DOI: 10.1016/j.gtc.2011.09.005]
 - 87 **Joly F**, Amiot A, Coffin B, Lavergne-Slove A, Messing B, Bouhnik Y. [Chronic intestinal pseudo-obstruction]. *Gastroenterol Clin Biol* 2006; **30**: 975-985 [PMID: 17075444 DOI: 10.1016/s0399-8320(06)73359-0]
 - 88 **Gu L**, Ding C, Tian H, Yang B, Zhang X, Hua Y, Zhu Y, Gong J, Zhu W, Li J, Li N. Serial Frozen Fecal Microbiota Transplantation in the Treatment of Chronic Intestinal Pseudo-obstruction: A Preliminary Study. *J Neurogastroenterol Motil* 2017; **23**: 289-297 [PMID: 27840368 DOI: 10.5056/jnm16074]
 - 89 **Ooms AH**, Verheij J, Hulst JM, Vlot J, van der Starre C, de Ridder L, de Krijger RR. Eosinophilic myenteric ganglionitis as a cause of chronic intestinal pseudo-obstruction. *Virchows Arch* 2012; **460**: 123-127 [PMID: 22173330 DOI: 10.1007/s00428-011-1183-x]
 - 90 **Fragulidis G**, Pantiora E, Michalaki V, Kontis E, Primetis E, Vezakis A, Polydorou A. Immune-

- related intestinal pseudo-obstruction associated with nivolumab treatment in a lung cancer patient. *J Oncol Pharm Pract* 2019; **25**: 487-491 [PMID: 29067858 DOI: 10.1177/1078155217738325]
- 91 **Forchielli ML**, Young MC, Flores AF, Richardson D, Lo CW. Immune deficiencies in chronic intestinal pseudo-obstruction. *Acta Paediatr* 1997; **86**: 1077-1081 [PMID: 9350888 DOI: 10.1111/j.1651-2227.1997.tb14811.x]
 - 92 **Halter JP**, Michael W, Schüpbach M, Mandel H, Casali C, Orchard K, Collin M, Valcarcel D, Rovelli A, Filosto M, Dotti MT, Marotta G, Barba P, Accarino A, Ferra C, Illa I, Beguin Y, Bakker JA, Boelens JJ, de Coe IF, Fay K, Sue CM, Nachbaur D, Zoller H, Sobreira C, Pinto Simoes B, Hammans SR, Savage D, Marti R, Chinnery PF, Elhasid R, Gratwohl A, Hirano M. Allogeneic haematopoietic stem cell transplantation for mitochondrial neurogastrointestinal encephalomyopathy. *Brain* 2015; **138**: 2847-2858 [PMID: 26264513 DOI: 10.1093/brain/awv226]
 - 93 **Bond GJ**, Reyes JD. Intestinal transplantation for total/near-total aganglionosis and intestinal pseudo-obstruction. *Semin Pediatr Surg* 2004; **13**: 286-292 [PMID: 15660322 DOI: 10.1053/j.sempedsurg.2004.10.016]
 - 94 **Lao OB**, Healey PJ, Perkins JD, Horslen S, Reyes JD, Goldin AB. Outcomes in children after intestinal transplant. *Pediatrics* 2010; **125**: e550-e558 [PMID: 20142294 DOI: 10.1542/peds.2009-1713]
 - 95 **Nakamura H**, Henderson D, Puri P. A meta-analysis of clinical outcome of intestinal transplantation in patients with total intestinal aganglionosis. *Pediatr Surg Int* 2017; **33**: 837-841 [PMID: 28600659 DOI: 10.1007/s00383-017-4107-2]
 - 96 **Schwankovsky L**, Mousa H, Rowhani A, DI Lorenzo C, Hyman PE. Quality of life outcomes in congenital chronic intestinal pseudo-obstruction. *Dig Dis Sci* 2002; **47**: 1965-1968 [PMID: 12353838 DOI: 10.1023/a:1019644022606]
 - 97 **Lauro A**, Zanfi C, Pellegrini S, Catena F, Cescon M, Cautero N, Stanghellini V, Pironi L, Pinna AD. Isolated intestinal transplant for chronic intestinal pseudo-obstruction in adults: long-term outcome. *Transplant Proc* 2013; **45**: 3351-3355 [PMID: 24182815 DOI: 10.1016/j.transproceed.2013.06.014]
 - 98 **Tzakis AG**, Kato T, Levi DM, Defaria W, Selvaggi G, Weppler D, Nishida S, Moon J, Madariaga JR, David AI, Gaynor JJ, Thompson J, Hernandez E, Martinez E, Cantwell GP, Augenstein JS, Gyamfi A, Pretto EA, Dowdy L, Tryphonopoulos P, Ruiz P. 100 multivisceral transplants at a single center. *Ann Surg* 2005; **242**: 480-90; discussion 491-3 [PMID: 16192808 DOI: 10.1097/01.sla.0000183347.61361.7a]
 - 99 **Kirby DF**, Raheem SA, Corrigan ML. Nutritional Interventions in Chronic Intestinal Pseudoobstruction. *Gastroenterol Clin North Am* 2018; **47**: 209-218 [PMID: 29413013 DOI: 10.1016/j.gtc.2017.09.005]
 - 100 **Minami T**, Nishibayashi H, Shinomura Y, Matsuzawa Y. Effects of erythromycin in chronic idiopathic intestinal pseudo-obstruction. *J Gastroenterol* 1996; **31**: 855-859 [PMID: 9027652 DOI: 10.1007/BF02358615]
 - 101 **Billiauw L**, Corcos O, Joly F. Dysmotility disorders: a nutritional approach. *Curr Opin Clin Nutr Metab Care* 2014; **17**: 483-488 [PMID: 25023191 DOI: 10.1097/MCO.0000000000000095]
 - 102 **Sutton DH**, Harrell SP, Wo JM. Diagnosis and management of adult patients with chronic intestinal pseudoobstruction. *Nutr Clin Pract* 2006; **21**: 16-22 [PMID: 16439766 DOI: 10.1177/011542650602100116]
 - 103 **Camilleri M**, Phillips SF. Acute and chronic intestinal pseudo-obstruction. *Adv Intern Med* 1991; **36**: 287-306 [PMID: 2024582]
 - 104 **Lehmann S**, Ferrie S, Carey S. Nutrition Management in Patients With Chronic Gastrointestinal Motility Disorders: A Systematic Literature Review. *Nutr Clin Pract* 2020; **35**: 219-230 [PMID: 30989698 DOI: 10.1002/ncp.10273]
 - 105 **Garipey CE**, Mousa H. Clinical management of motility disorders in children. *Semin Pediatr Surg* 2009; **18**: 224-238 [PMID: 19782304 DOI: 10.1053/j.sempedsurg.2009.07.004]
 - 106 **Gabbard SL**, Lacy BE. Chronic intestinal pseudo-obstruction. *Nutr Clin Pract* 2013; **28**: 307-316 [PMID: 23612903 DOI: 10.1177/0884533613485904]
 - 107 **Vasant DH**, Pironi L, Barbara G, Bozzetti F, Cuerda C, Joly F, Mundi M, Paine P, Staun M, Szczepanek K, Van Gossum A, Wanten G, Lal S. An international survey on clinicians' perspectives on the diagnosis and management of chronic intestinal pseudo-obstruction and enteric dysmotility. *Neurogastroenterol Motil* 2020; e13937 [PMID: 32696607 DOI: 10.1111/nmo.13937]
 - 108 **Lauro A**, Zanfi C, Dazzi A, di Gioia P, Stanghellini V, Pironi L, Ercolani G, Gaudio MD, Ravaioli M, Faenza S, di Simone M, Pinna AD. Disease-related intestinal transplant in adults: results from a single center. *Transplant Proc* 2014; **46**: 245-248 [PMID: 24507060 DOI: 10.1016/j.transproceed.2013.08.110]
 - 109 **Krasaelap A**, Kovacic K, Goday PS. Nutrition Management in Pediatric Gastrointestinal Motility Disorders. *Nutr Clin Pract* 2020; **35**: 265-272 [PMID: 31321821 DOI: 10.1002/ncp.10319]
 - 110 **Pironi L**, Arends J, Bozzetti F, Cuerda C, Gillanders L, Jeppesen PB, Joly F, Kelly D, Lal S, Staun M, Szczepanek K, Van Gossum A, Wanten G, Schneider SM. Home Artificial Nutrition & Chronic

- Intestinal Failure Special Interest Group of ESPEN. ESPEN guidelines on chronic intestinal failure in adults. *Clin Nutr* 2016; **35**: 247-307 [PMID: 26944585 DOI: 10.1016/j.clnu.2016.01.020]
- 111 **Scolapio JS**, Ukleja A, Bouras EP, Romano M. Nutritional management of chronic intestinal pseudo-obstruction. *J Clin Gastroenterol* 1999; **28**: 306-312 [PMID: 10372926 DOI: 10.1097/00004836-199906000-00005]
 - 112 **Faure C**, Goulet O, Ategbo S, Breton A, Tounian P, Ginies JL, Roquelaure B, Despres C, Scaillon M, Maurage C, Paquot I, Hermier M, De Napoli S, Dabadie A, Huet F, Baudon JJ, Larchet M. Chronic intestinal pseudoobstruction syndrome: clinical analysis, outcome, and prognosis in 105 children. French-Speaking Group of Pediatric Gastroenterology. *Dig Dis Sci* 1999; **44**: 953-959 [PMID: 10235603 DOI: 10.1023/a:1026656513463]
 - 113 **Ukleja A**, Romano MM. Complications of parenteral nutrition. *Gastroenterol Clin North Am* 2007; **36**: 23-46, v [PMID: 17472873 DOI: 10.1016/j.gtc.2007.01.009]
 - 114 **Boybeyi Türer Ö**, Soyer T, Özen H, Arslan UE, Karnak İ, Tanyel FC. Challenges in management and prognosis of pediatric intestinal pseudo-obstruction. *Turk J Gastroenterol* 2020; **31**: 596-602 [PMID: 32915148 DOI: 10.5152/tjg.2020.19233]
 - 115 **Di Lorenzo C**, Flores AF, Buie T, Hyman PE. Intestinal motility and jejunal feeding in children with chronic intestinal pseudo-obstruction. *Gastroenterology* 1995; **108**: 1379-1385 [PMID: 7729629 DOI: 10.1016/0016-5085(95)90685-1]
 - 116 **Hunter MF**, Peters H, Salemi R, Thorburn D, Mackay MT. Alpers syndrome with mutations in POLG: clinical and investigative features. *Pediatr Neurol* 2011; **45**: 311-318 [PMID: 22000311 DOI: 10.1016/j.pediatrneurol.2011.07.008]
 - 117 **Deglincerti A**, De Giorgio R, Cefle K, Devoto M, Pippucci T, Castegnaro G, Panza E, Barbara G, Cogliandro RF, Mungan Z, Palanduz S, Corinaldesi R, Romeo G, Seri M, Stanghellini V. A novel locus for syndromic chronic idiopathic intestinal pseudo-obstruction maps to chromosome 8q23-q24. *Eur J Hum Genet* 2007; **15**: 889-897 [PMID: 17487221 DOI: 10.1038/sj.ejhg.5201844]
 - 118 **Gargiulo A**, Auricchio R, Barone MV, Cotugno G, Reardon W, Milla PJ, Ballabio A, Ciccodicola A, Auricchio A. Filamin A is mutated in X-linked chronic idiopathic intestinal pseudo-obstruction with central nervous system involvement. *Am J Hum Genet* 2007; **80**: 751-758 [PMID: 17357080 DOI: 10.1086/513321]



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