Chronic Idiopathic Intestinal Pseudo-Obstruction (CIIP)

42

Mitsuru Muto

42.1 Disease Concept

Intestinal pseudo-obstruction was first described in 1958 by Dudley et al. [1]. The reported case presented with serious ileus without mechanical obstruction. Faulk et al. proposed the term chronic intestinal pseudo-obstruction (CIPO) in 1978 [2]. CIPO is a functional motility disorder presenting with repetitive or continuous nonmechanical bowel obstructive symptoms, such as abdominal distension, nausea and vomiting, abdominal pain, and intestinal dilatation. Historically, CIPO in children has been variously referred to as chronic intestinal pseudo-obstruction (CIP), chronic intestinal pseudo-obstruction syndrome (CIPS or CIPOS), pseudo-Hirschsprung's disease, and chronic adynamic ileus [3–7]. This suggests that the epidemiological and clinical features of CIPO in children have been unclear.

CIPO is classified into three types: primary type, caused by gastrointestinal lesions; secondary type, associated with systemic illness or drugs; and "idiopathic" type with unknown etiology. CIPO is recognized as chronic idiopathic intestinal pseudo-obstruction (CIIP) when a conventional histological examination, such as hematoxylin and eosin staining, fails to show a meaningful pathology [3, 8].

As primary CIPO includes Hirschsprung's disease (aganglionosis) and allied Hirschsprung's diseases (except CIIP) (refer to other chapters), a diagnosis of childhood CIIP requires differentiation from these diseases. It is also important to differentiate secondary CIPO in the diagnosis of adult CIIP.

CIIP is characterized as a rare and intractable disease with an unknown etiology for which an effective treatment method has not yet been established [9]; high-quality evidence concerning its treatment is extremely lacking. This chapter mainly describes CIIP that develops in childhood based on a nationwide survey conducted in Japan and newly released clinical practice guidelines for allied Hirschsprung's disease by Tomoaki Taguchi's project group on research for "establishing guidelines for rare and intractable gastrointestinal diseases, spanning childhood to the transition phase." Supporting articles include journals in Japanese.

42.2 The Diagnosis

In this section, we will describe the diagnostic criteria for childhood CIIP newly established in Japan (Table 42.1).

The recognition of prolonged obstructive symptoms and exclusion of mechanical obstruction are key for the diagnosis of CIIP. The disease is considered to be "chronic" if the functional obstructive symptoms appear during the neonatal period and persist for the first 2 months of life or if it appears after the neonatal period and persists for longer than 6 months [3]. When no pathological abnormalities in the ganglion plexus of the affected intestine are exhibited, the disease is described as "idiopathic." "Pseudo-obstruction" denotes signs and symptoms resembling a physical obstruction to the luminal flow, including radiographic documentation of dilated bowel with air-fluid levels, in the absence of a true mechanical obstruction.

The duration of symptoms and the bowel obstructive conditions are determined from the clinical history and physical examination findings. The presence of intestinal dilatation, air-fluid level formation, and the absence of mechanical obstruction are confirmed by plain abdominal X-ray, computed tomography (CT), magnetic resonance imaging (MRI), or similar modalities [10, 11]. For neonates, radiological examinations in a standing or lateral decubitus position may be difficult, considering patient's condition, so the confirmation of air-fluid levels by plain abdominal X-ray is not necessarily required in neonatal cases. In the diagnosis of childhood CIIP, the location of dilated segments, the degree

M. Muto (✉)
Department of Neonatology, Perinatal Medical Center, Kagoshima City Hospital, Kagoshima, Japan

Department of Pediatric Surgery, Research Field in Medicine and Health Sciences, Medical and Dental Sciences Area, Research and Education Assembly, Kagoshima University, Kagoshima, Japan
e-mail: mitsuru@m3.kufm.kagoshima-u.ac.jp

T. Taguchi et al. (eds.), *Hirschsprung's Disease and the Allied Disorders*, https://doi.org/10.1007/978-981-13-3606-5_42

Table 42.1 Diagnostic criteria for chronic idiopathic intestinal pseudo-obstruction

The following seven items need to be satisfied:
1. Persistent or repetitive development over a long period of time of serious bowel obstructive symptoms that may require hospitalization, such as abdominal distension, nausea and vomiting, abdominal pain, etc.
2. Duration of symptoms is ≥2 months for neonatal onset and ≥6 months for onset in infancy or later
3. Gastrointestinal dilatation and air-fluid level noted on diagnostic imaging[a]
4. No lesions mechanically blocking the gastrointestinal tract
5. No pathological abnormalities in the nerve plexuses on HE staining obtained by an intestinal full-thickness biopsy[b]
6. Megacystis-microcolon-intestinal hypoperistalsis syndrome and segmental dilatation of intestine excluded
7. Secondary chronic intestinal pseudo-obstruction excluded[c]

[a]Confirmation of the air-fluid level on plain abdominal radiography in a standing position is not necessarily required for neonates.
[b]For adults, when an intestinal full-thickness biopsy is unobtainable, a characteristic peristalsis disorder should be confirmed by manometry or cine-MRI.
[c]Table 42.2 shows the secondary chronic intestinal pseudo-obstructions to be excluded.

of peristaltic disorder, and the presence of nonmechanical obstruction can be detected based on the gastrointestinal gas pattern on plain abdominal X-ray and gastrointestinal series. An intestinal full-thickness biopsy is indispensable for the definitive diagnosis of idiopathy and exclusion of other allied Hirschsprung's diseases, such as isolated hypoganglionosis and immaturity of ganglia [11, 12]. Its idiopathy is confirmed when conventional histological examination, such as hematoxylin and eosin staining, fails to show a meaningful pathology [8].

In adult cases of CIIP, it is important to differentiate true mechanical obstructions caused by neoplastic lesions, inflammation, and adhesion. In addition, the exclusion of secondary pseudo-obstructions is emphasized in the diagnosis of idiopathy. The secondary pseudo-obstructions to be differentiated are listed in Table 42.2. Generally, a full-thickness biopsy is not conducted in adult cases as it is for children. Cine-MRI and intestinal manometry are useful for assessing peristaltic disorders present in CIIP, and these two modalities are used instead of a full-thickness biopsy in adults [8, 13–15].

42.3 Clinical Manifestation

In February 2012, we conducted a nationwide survey in Japan for an assessment of epidemiological and clinical features of CIPO among children, involving facilities represented by members of the Japanese Society of Pediatric Surgeons; the Japanese Society for Pediatric Gastroenterology, Hepatology and Nutrition; and the Japanese Study Group of Pediatric Constipation [12]. A total of 92 responses were collected from 47 pediatric facilities, and 62 cases were diagnosed as pediatric CIPO. Considering the population of Japanese children younger than 15 years of age (16.7 million), the esti-

Table 42.2 Secondary chronic intestinal pseudo-obstruction

1. Gastrointestinal smooth muscle-related diseases
Systemic sclerosis
Dermatomyositis
Multiple myositis
Systemic lupus erythematosus
Mixed connective tissue disease (MCTD)
Ehlers-Danlos syndrome
Muscular dystrophy
Amyloidosis
Small bowel-based lymphoid infiltration
Brown bowel syndrome (Ceroidosis)
Mitochondrial encephalomyopathy
2. Gastrointestinal nerves-related diseases
Familial dysautonomia
Primary dysautonomia
Diabetic neuropathy
Myotonic dystrophy
Pseudo-obstruction after infection, such as EB virus, Herpes Zoster virus, and Rota virus
3. Endocrine diseases
Hypothyroidism
Hypoparathyroidism
Phaeochromocytoma
4. Metabolic diseases
Uremia
Porphyria
Serious electrolytes abnormality (K^+, Ca^{2+}, Mg^{2+})
5. Others
Celiac disease
Kawasaki disease
Eosinophilic enteritis
Paraneoplastic pseudo-obstruction
Mesenteric vein thrombosis
Side reactions to radiotherapy
Angioedema
Intestinal tuberculosis
Crohn's disease
Chagas disease
Paralytic ileus resulting from injury, after gastrointestinal surgery, intraperitoneal inflammation, etc.
Ogilvie syndrome
6. Drug-induced diseases
Antidepressant
Antianxiety drug
Anthraquinone-based laxative
Phenothiazine drugs
Vinca alkaloid
Anticholinergic drug
Opioid
Ca channel blocker
Verapamil

mated prevalence of pediatric CIPO was determined to be 3.7 per one million children. Pediatric CIPO therefore seems to be an extremely rare disease. Among these 62 cases, 49 (23 males, 26 females) met the diagnostic criteria of CIIP. The mean age of the patients was 12.1 years (median, 8 years; range, 0–43 years) at the time of the survey.

According to a previous nationwide survey conducted in the Japanese adult population, the number of patients with CIPO peaks at 40–50 years of age [16]. In contrast, in the pediatric population, CIPO as well as CIIP develops during the neonatal period in more than half of cases (Fig. 42.1).

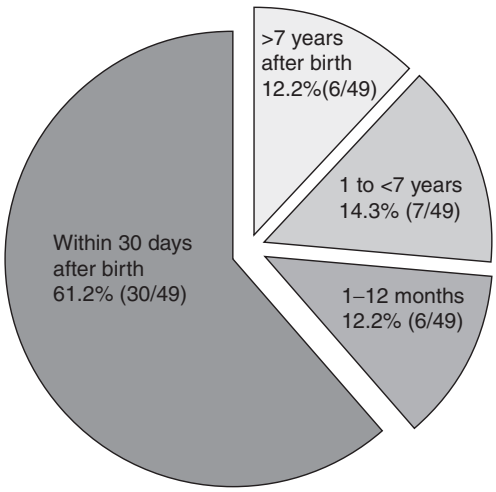

Fig. 42.1 Disease onset. More than 60% of patients developed CIIP in the neonatal period in Japan

Familial accumulation was noted in two families, and four female patients had familial incidence. A set of twins, both with megacystis (without presence of microcolon), developed CIIP during neonatal period, while two sisters, both with galactosemia, developed CIIP at school age.

The major initial symptoms are abdominal distension and vomiting for neonatal-onset cases and abdominal distension, vomiting, and chronic constipation for postneonatal-onset cases (Fig. 42.2). Peristalsis disorders may occur at single or multiple sites of the gastrointestinal tract. Dilated intestines are frequently observed in the small bowel and colon. In the majority of CIIP cases, remission and exacerbation of those pseudo-obstructive symptoms may be repeated with progressing conditions (Fig. 42.3). Repeated long-term hospitalization is therefore sometimes required. Oral intake was restricted in three-fourths of the CIIP patients, and more than half were dependent on parenteral nutrition in our former survey (Fig. 42.4).

We formulated severity criteria based on factors that impair the quality of a patient's daily life (Table 42.3). According to these criteria, 43 out of 49 (87.8%) CIIP children were assessed as severe cases.

CIIP is summarized as a rare, serious, and intractable disease.

42.4 Therapeutic Interventions

Treatment may start with conservative treatment, such as drug therapy and intravenous/enteral nutrition, and shift to invasive treatments, such as decompression by tubing or

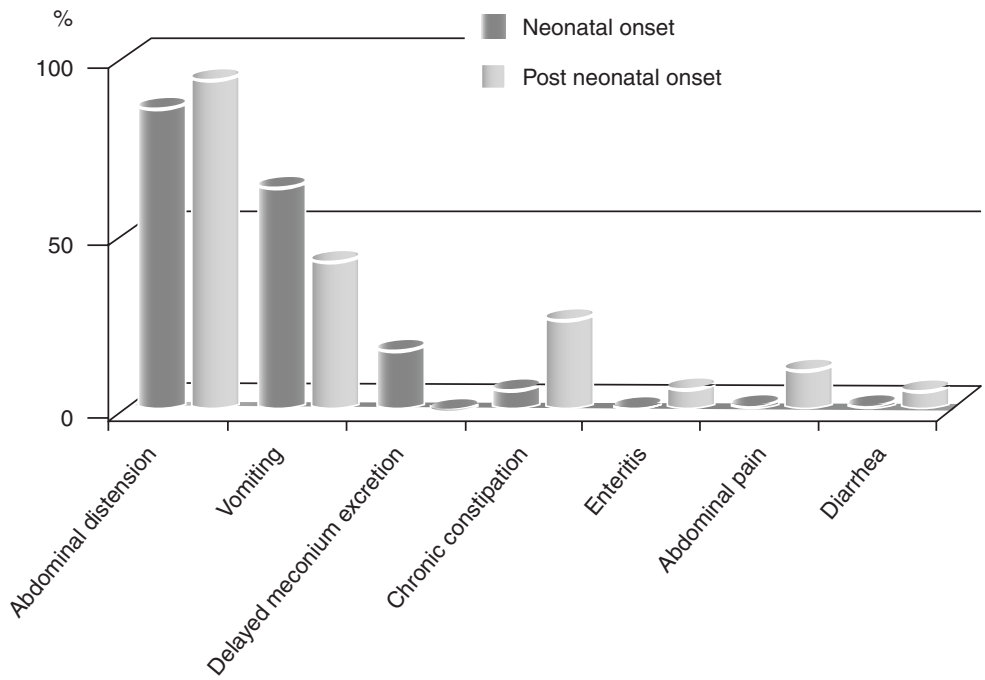

Fig. 42.2 Initial symptoms and signs (multiple answers allowed)

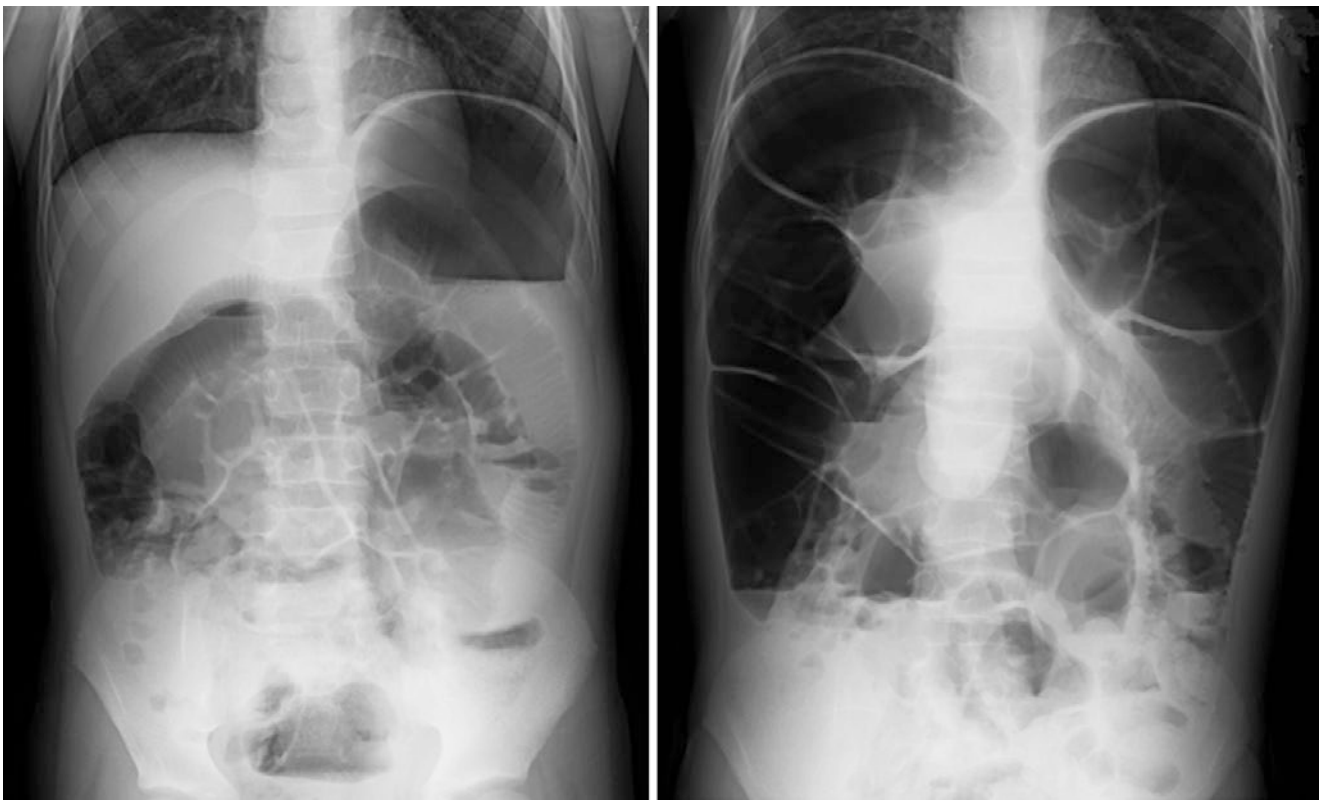

Fig. 42.3 Plain abdominal X-ray of a childhood with CIIP. At 7 years of age (left), intestinal dilatation and air-fluid were present. At 17 years of age (right), the dilatation had progressed, and free air was observed under the diaphragm

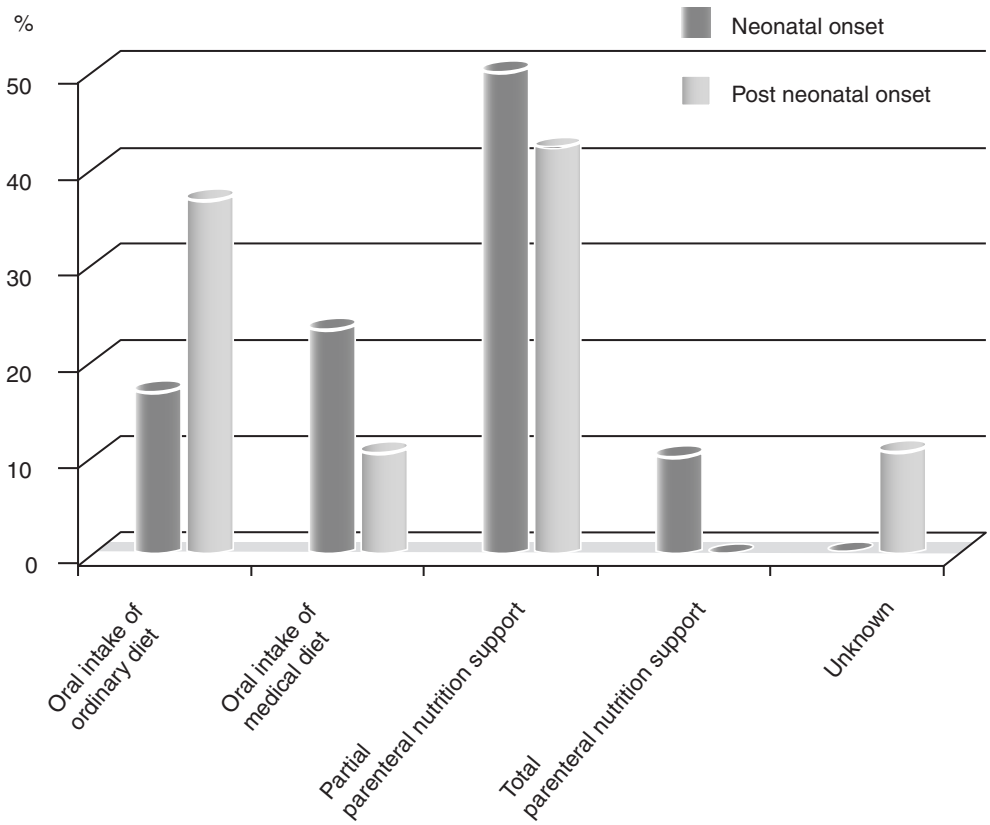

Fig. 42.4 Nutritional management. Only a quarter (24.5%; 12/49) of the patients were receiving a normal oral diet independent of any nutritional support. Half (53.1%; 26/49) of the patients depended on parenteral nutritional support

Table 42.3 Severity criteria

Severe case is defined as one whose daily life is significantly impaired due to bowel obstructive symptoms, such as abdominal distension, nausea and vomiting, and abdominal pain, and when at least one of the following three items is met:
1. Parenteral nutrition is required
2. Enteral nutrition is required
3. Continuous gastrointestinal decompression is required[a]

[a]Gastrointestinal decompression refers to the drainage of intestinal contents through enterostomy, gastrostomy, nasogastric tube, ileus tube, transanal tube, etc.

enterostomy, as the condition progresses. However, no treatment modality has been clearly shown to be effective in relieving symptoms.

42.4.1 Drug Therapy

Some drugs are used to control CIIP, including prokinetic agents, Chinese medicine (dai-kenchu-tou), probiotics, antibiotics, laxatives, and antidiarrheals. However, few randomized control trials (RCTs) or case series have provided supporting evidence, and the available articles are mostly case reports. There are no recommended medications [17].

Attempts have been made to improve pseudo-obstructive symptoms with prokinetic agents; one case series examined the efficacy of prucalopride [11], and one cross-sectional study examined the efficacy of cisapride [18]. Some case reports have suggested the usefulness of cisapride, the administration of which resulted in an increased intake of enteral nutrients and a reduction in the intestinal transit time [19–21]. However, other case reports have found this agent to be ineffective [22–24]. The effectiveness of mosapride for CIIP has not yet been reported.

Two case reports have described the usefulness of dai-kenchu-tou, a Chinese medicine, in improving gastrointestinal motility and bowel obstructive symptoms [25, 26]; however, one case report conversely found it to be ineffective [27].

Two case reports have described an increased intake of enteral nutrition and reduced incidence of enteritis treating with probiotics [28, 29]. No adverse events associated with probiotics have been reported, but evidence supporting its effectiveness is not sufficient.

Erythromycin is sometimes administered to enhance intestinal motility, and some case reports have shown its effects on increasing the enteral nutrition intake [27, 30]. One case report described the improvement of pseudo-obstruction with polymyxin B [31]. No adverse events have been reported with the administration of antibiotics for CIIP, but evidence supporting their efficacy is lacking at present.

Regarding other drugs, one case report found laxatives to be effective [20], and two case reports found that prostaglandin improved bowel obstructive symptoms [31, 32]. Again, however, the effectiveness of those agents remains unclear. Buprenorphine, a weak opioid, was reported as an agent for relieving abdominal pain [14]. Although the evidence is not sufficient, buprenorphine may be useful for relieving symptoms of abdominal pain associated with CIIP.

42.4.2 Nutritional Therapy

CIIP is a disease that requires repeated, prolonged fasting and central venous nutrition management due to enteritis caused by stagnation of intestinal contents. Advances in the management of optimal parenteral/enteral nutrition support may prolong the survival of patients [14, 33].

Since CIIP may have a long duration of symptoms, special attention needs to be paid to the life-threatening complications related to long-term parenteral nutrition, such as trace element deficiency, electrolyte disturbance, parenteral nutrition-associated liver disease (PNALD), and catheter-related blood stream infections (CRBSIs). Of the 49 cases in our survey, 18 (36.7%) had PNALD, and CRBSIs were seen in 10 (20.4%).

Semi-digested nutrients, the concomitant use of semi-digested nutrients and a low-residue diet, and digestive nutrients are reported to be useful for enteral nutrition in patients with a functional ileus [21]. However, enteral nutrition may not be able to be administered in cases with aggravated symptoms. Further investigation into what kinds of enteral nutrients are most useful is needed.

42.4.3 Gastrointestinal Decompression

Appropriate gastrointestinal decompression should be considered on a case-by-case basis. Intermittent decompression via enteric tube may be effective in some CIIP cases, and enterostomy may be effective in other cases. Gastrointestinal decompression may enable enteral feeding and encourage growth and long-term survival. In our nationwide survey, decompression of the dilated tracts was performed in 39 CIIP children (79.6%, 39/49), 28 of whom (71.8%, 28/39) had permanent enterostomas. Of these 28 cases, five (17.9%) were able to receive oral intake of an ordinary diet.

Nasogastric tube decompression for CIIP has been reported to be effective in the short term. Regarding the nature of CIIP, once the bowel pseudo-obstruction symptoms subside, they may relapse again [34]. The placement of an ileus tube may improve abdominal distension and abdominal pain remarkably, but its long-term efficacy is not clear [35]. There have been some cases in which oral intake was enabled by intestinal lavage and continuous decompression through enterostomy [36]. Antegrade continence enema may be sug-

gested to improve defecation and abdominal distension [26]. Although temporary efficacy has been reported with both jejunostomy and ileostomy, colonostomy has not been shown to be effective in any cases and has conversely been reported not to have improved bowel obstruction symptoms at all [37]. No side effects or adverse events accompanying decompression therapy have been reported.

42.4.4 Radical Surgical Treatment

When gastrointestinal decompression is not effective, the risk of intestinal perforation or enteritis should be kept in mind. Unfortunately, however, even if the dilated segments are resected, obstructive symptoms may relapse due to dysfunction in the remaining intestine. Intestinal resection is not believed to improve the pseudo-obstructive symptoms. Avoiding multiple surgery is recommended in the treatment of CIIP [9, 16].

In cases of duodenal dilatation, duodenojejunostomy is suggested to improve the intestinal transit of the dilated segment. It may relieve the pseudo-obstructive symptoms to some extent [38].

In our nationwide survey, 13 out of 49 patients (26.5%) underwent intestinal resection. However, less than one-quarter (3/13, 23.1%) were able to receive oral intake of an ordinary diet; the rest continued to depend on parenteral nutrition or required enteral nutrition with a medical diet. A tendency toward undergoing multiple surgeries was seen in the group treated with intestinal resection compared with the group treated without intestinal resection (average number of surgical interventions: 3.85 times vs. 2.33 times, $p = 0.047$). Attempts to improve pseudo-obstruction by resection of the dilated intestine, including ileocecal resection and colectomy, failed in several studies [24, 30, 39]. No reports have shown the efficacy of intestinal resection with clear evidence.

42.4.5 Small Bowel Transplantation

Small bowel transplantation may be indicated as a final measure when conservative treatments become ineffective due to complications, when patients have suffered from intolerable pain for a long period of time, or when repeated catheter-related infections lead to the loss of central venous access and progression of hepatopathy. Since CIIP is often characterized by disorder of gastric emptying, the impaired native gastric outlet function needs to be considered when small bowel transplantation is planned and performed.

Multivisceral transplantation is generally performed [40, 41], and isolated small bowel transplantation plus partial gastrectomy with graft-gastric or graft-duodenal anastomoses may be another option [42].

In Japan, isolated small bowel transplantation for CIIP has been performed in three cases. In two of these cases, two anastomoses were performed at the oral side to add graft-duodenal or graft-jejunal anastomosis to graft-gastric anastomosis. In both cases, however, oral intake was not easily established. Only one out of the three cases has survived.

42.5 The Prognosis of Chronic Idiopathic Intestinal Pseudo-Obstruction

An estimated 10–25% children with CIPO died before adulthood in the past quarter century [4, 5]. Advances in the management of intestinal failure, including optimal nutrition support and improvements in the management of sepsis, may prolong the survival of CIIP patients. In our survey in Japan, only three children with CIIP died from enteritis or sepsis, resulting in a survival rate of 93.9% (46/49). The number of cases transitioning from childhood to adult is likely to increase in the future [12].

In clinical practice, repeated hospitalization is sometimes required for patients with CIIP due to exacerbation of pseudo-obstructive symptoms. Even for outpatient cases, the requirement of intravenous nutrition support and enterostomy itself may significantly limit patients' daily life. While the prognosis of CIIP with respect to the survival is good, this outcome is deemed unsatisfactory given the restriction of patients' long-term quality of daily life [43].

References

1. Dudley HA, Sinclair IS, IF ML, TJ MN, Newsam JE. Intestinal pseudo-obstruction. J R Coll Surg Edinb. 1958;3:206–17.
2. Faulk DL, Anuras S, Christensen J. Chronic intestinal pseudoobstruction. Gastroenterology. 1978;74:922–31.
3. Rudolph CD, Hyman PE, Altschuler SM, Christensen J, Colletti RB, Cucchiara S, et al. Diagnosis and treatment of chronic intestinal pseudo-obstruction in children: report of consensus workshop. J Pediatr Gastroenterol Nutr. 1997;24:102–12.
4. Mousa H, Hyman PE, Cocjin J, Flores AF, Di Lorenzo C. Long-term outcome of congenital intestinal pseudoobstruction. Dig Dis Sci. 2002;47:2298–305.
5. Faure C, Goulet O, Ategbo S, Breton A, Tounian P, Ginies JL, et al. Chronic intestinal pseudoobstruction syndrome: clinical analysis, outcome and prognosis in 105 children. French-Speaking Group of Pediatric Gastroenterology. Dig Dis Sci. 1999;44:953–9.
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;7:323–32.
7. Goulet O, Jobert-Giraud A, Michel JL, Jaubert F, Lortat-Jacob S, Colomb V, et al. Chronic intestinal pseudo-obstruction syndrome in pediatric patients. Eur J Pediatr Surg. 1999;9:83–9.

8. Stanghellini V, Cogliandro RF, De Giorgio R, Barbara G, Morselli-Labate AM, Cogliandro L, et al. Natural history of chronic idiopathic intestinal pseudo-obstruction in adults: a single center study. Clin Gastroenterol Hepatol. 2005;3:449–58.
9. Muto M, Matsufuji H, Muranaga F, Tomomasa T, Nakajima A, Taguchi T. Data mined distinctive features of pediatric chronic idiopathic intestinal pseudo-obstruction in Japan. Pediat Ther. 2015;5:1–6. https://doi.org/10.4172/2161-0665.1000236.
10. Byrne WJ, Cipel L, Ament ME, Gyepes MT. Chronic idiopathic intestinal pseudo-obstruction syndrome. Radiologic signs in children with emphasis on differentiation from mechanical obstruction. Diagn Imaging. 1981;50:294–304.
11. Muñoz-Yagüe MT, Solís-Muñoz P, Salces I, Ballestín C, Colina F, Ibarrola C, et al. Chronic intestinal pseudo-obstruction: a diagnosis to be considered. Rev Esp Enferm Dig. 2009;101:336–42.
12. Muto M, Matsufuji H, Tomomasa T, Nakajima A, Kawahara H, Ida S, et al. Pediatric chronic intestinal pseudo-obstruction is a rare, serious, and intractable disease: a report of a nationwide survey in Japan. J Pediatr Surg. 2014;49:1799–803.
13. Tomomasa T. Findings from gastroduodenal manometry in Chronic idiopathic intestinal pseudo-obstruction - especially on phase 3. J Smooth Muscle Res. 1993;29:204–6.. (Article in Japanese).
14. Muñoz-Yagüe MT, Marín JC, Colina F, Ibarrola C, López-Alonso G, Martín MA, et al. Chronic primary intestinal pseudo-obstruction from visceral myopathy. Rev Esp Enferm Dig. 2006;98:292–302.
15. Irtan S, Bellaïche M, Brasher C, El Ghoneimi A, Cézard JP, Bonnard A. Stomal prolapse in children with chronic intestinal pseudoobstruction: a frequent complication? J Pediatr Surg. 2010;45:2234–7.
16. 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;23:288–94.
17. Hashizume N, Yagi M, Ushijima K, Seki Y, Fukahori S, Muto M, et al. Pharmacotherapy for pediatric chronic intestinal pseudo-obstruction: nationwide survey in Japan. Pediatr Int. 2017;59:467–72.
18. Tanabe M, Takahashi H, Oonuma N, Yoshida H, Niibo K. Experience of chronic idiopathic intestinal pseudo-obstruction (CIIP) treated with cisapride. J Jpn Soc Pediatr Surg. 1991;27:48–55.. (Article in Japanese).
19. Horiuchi I, Ito H, Yamashita T, Kondo K, Eguchi T, Watanabe Y. A case of Chronic idiopathic intestinal pseudo-obstruction developed in neonatal period. J Jpn Soc Pediatr Surg. 1992;28:1359–63.. (Article in Japanese).
20. Miyauchi K, Ohshima Y, Shimizu M, Watanabe T, Yoshida K, Tomonaga T. A case of Chronic idiopathic intestinal pseudo-obstruction and investigation on reported cases in Japan. J Jpn Soc Gastroenterol. 1991;88:1359–63.. (Article in Japanese).
21. Ament ME, Vargas J. Diagnosis and management of chronic intestinal pseudo-obstruction syndromes in infancy and childhood. Arq Gastroenterol. 1988;25:157–65.
22. Suita R, Sai H, Izumi H. An autopsy case of Chronic idiopathic intestinal pseudo-obstruction syndrome (CIIPS). Jpn J Pediatr. 1983;36:135–42.. (Article in Japanese).
23. Yahano S, Furuya S, Kadowaki H. Clinical course of 8-year-old girl with CIIP and problems of long-term TPN support. Jpn J Pediatr Surg. 1983;15:1129–33.. (Article in Japanese).
24. Ozawa M, Yamada R, Oohama M. Two cases of chronic idiopathic intestinal pseudo-obstruction syndrome (CIIPS). Jpn J Pediatr Surg. 1983;15:1139–46.. (Article in Japanese).
25. Kubo M, Yanai T. Experiences of treatment with Dai-kenchu-tou for various kinds of ileus. Prog Med. 1997;17:2547–9.. (Article in Japanese).
26. Kubota A, Kawahara H, Hasegawa T, Okuyama H, Uehara S, Ishikawa N, et al. Surgical intervention for chronic constipation. Jpn J Pediatr Surg. 2008;40:226–34.. (Article in Japanese).
27. Hirakawa H, Ueno S, Matuda H, Hinoki T, Kato Y. Effect of the herbal medicine dai-kenchu-to on gastrointestinal motility in patients with megacystis-microcolon-intestinal hypoperistalsis syndrome (MMIHS) and chronic idiopathic intestinal pseudo-obstruction (CIIP): report of two cases. Tokai J Exp Clin Med. 2009;34:28–33.
28. Kiyohara Y, Nakanaga M, Takahashi T, Ida S, Kubota A. The efficacy of symbiotics for allied Hirschsprung's disease. J JSPEN. 2010;25:935–9.. (Article in Japanese).
29. Onishi S, Wada H, Tamai S, Funado M. A case of CIIPS who was struggling to treat weight gain and intestinal infection. Kinki Shinseiji Kenkyukai Kaishi. 2005;14:24–8.. (Article in Japanese).
30. Dalgiç B, Sari S, Doğan I, Unal S. Chronic intestinal pseudoobstruction: report of four pediatric patients. Turk J Gastroenterol. 2005;16:93–7.
31. Yoshida T, Murai K, Sugimoto S. Two cases considered to be Chronic idiopathic intestinal pseudo-obstruction. Jpn J Clin Radiol. 1997;31:335–8.. (Article in Japanese).
32. Michiura J, Katsumi M, Kawano N, Aoki Y, Taniguchi K, Sakaguchi M, et al. A case of Chronic idiopathic intestinal pseudo-obstruction with mitral valve divergence syndrome. Jpn J Gastroenterol Surg. 1988;21:1134–7.. (Article in Japanese).
33. Antonucci A, Fronzoni L, Cogliandro L, Cogliandro RF, Caputo C, De Giorgio R. Chronic intestinal pseudo-obstruction. World J Gastroenterol. 2008;14:2953–61.
34. Tachibana M, Nakazawa Y, Sakamoto Y. A case of Chronic idiopathic intestinal pseudo-obstruction; its complications and problems on long-term management. Gastroenterology. 1987;6:421–8.. (Article in Japanese).
35. Matsuura A, Kikuchi T, Ooshima T. A case of Chronic idiopathic intestinal pseudo-obstruction with transverse colonic axial torsion. J Abdom Emerg Med. 1996;16:1321–4.. (Article in Japanese).
36. Nishijima E, Anpuku M, Higashimoto Y, Muraji T, Tsugawa C. Treatment experience of Chronic idiopathic intestinal pseudo-obstruction (CIIPS) - complications and problems on nutritional management of 8 cases developed in infancy. J J Parent Enteral Nutr. 1992;14:1395–401.. (Article in Japanese).
37. Kubota A, Okuyama H, Takahashi T. Pathology and treatment of intestinal torsion in children. A case report of CIIPS caused by intestinal malrotation - consideration on the difference between CIIPS and MMIHS. Jpn J Pediatr Surg. 2005;37:824–31.. (Article in Japanese).
38. Amano Y, Nakahata R, Akahane T. Cases of siblings with Chronic idiopathic intestinal pseudoobstruction (CIIP). J Pediatr Pract. 1985;48:937–41.. (Article in Japanese).
39. Nozawa H, Hori T, Kubo K. An older child with chronic idiopathic intestinal pseudo-obstruction presenting chronic constipation – problems on diagnosis. Jpn J Pediatr Surg. 1983;15:1135–8.. (Article in Japanese).
40. Loinaz C, Mittal N, Kato T, Miller B, Rodriguez M, Tzakis A. Multivisceral transplantation for pediatric intestinal pseudo-obstruction: single center's experience of 16 cases. Transplant Proc. 2004;36:312–3.
41. Sigurdsson L, Reyes J, Kocoshis SA, Mazariegos G, Abu-Elmagd KM, Bueno J, et al. Intestinal transplantation in children with chronic intestinal pseudo-obstruction. Gut. 1999;45:570–4.
42. Lauro A, Zanfi C, Pellegrini S, Catena F, Cescon M, Cautero N, et al. Isolated intestinal transplant for chronic intestinal pseudo-obstruction in adults: long-term outcome. Transplant Proc. 2013;45:3351–5.
43. Taguchi T, Ieiri S, Miyoshi K, Kohashi K, Oda Y, Kubota A, et al. The incidence and outcome of allied disorders of Hirschsprung's disease in Japan: results from a nationwide survey. Asian J Surg. 2017;40:29–34.