Digestive Diseases and Sciences, Vol. 44, No. 5 (May 1999), pp. 953–959

Chronic Intestinal Pseudoobstruction Syndrome

Clinical Analysis, Outcome, and Prognosis in 105 Children

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Our aim was to collect a large number of cases to characterize clinical presentation, outcome, and prognosis of chronic intestinal pseusoobstruction in children. We conducted a retrospective multicenter study that included children treated for chronic intestinal pseusoobstruction defined as recurrent episodes of intestinal obstruction with no mechanical obstruction, excluding Hirschsprung's disease. In all, 105 children, 57 boys and 48 girls, were studied, including five familial forms. Prenatal diagnosis was made in 18 patients. Eighty patients were less than 12 months old at onset; the disease began at birth for 37 patients. The most frequent signs were abdominal distension, vomiting, and constipation. Megacystis was noted in myopathies (7 cases), neuropathies (10 cases) and unclassified forms (13 cases). For all but three cases (two patients with CMV infection, one with Munchhausen-by-proxy syndrome), the associated diseases and disorders could not account for chronic intestinal pseusoobstruction as a secondary disorder. At least one full-thickness biopsy from the digestive tract was studied for 99 patients. The diagnosis recorded was visceral neuropathy in 58 cases, visceral myopathy in 17 cases, and uncertain or normal biopsy results in 24 cases. Seventy-eight children were fed intravenously, and only 18 were able to be fed orally throughout their illness. Seventy-one patients underwent surgery during their illness, and 217 surgical procedures, a mean of 3 per patient, were performed. Ostomy was the most performed procedure. Follow-up continued in 89 patients for 3 months to 16 years (mean 85 months). Forty-two patients were still fed by parenteral (39 patients) or enteral nutrition (3 patients) at the time of the study. Eleven patients died between the age of 1 month and 14 years 7 months.

KEY WORDS: chronic intestinal pseudoobstruction; children; surgery; nutritional support.

Chronic intestinal pseusoobstruction (CIP) is a rare disorder in which impaired intestinal motility leads to signs and symptoms of intestinal obstruction in the absence of a mechanical obstructive lesion (1). Knowledge and treatment of this heterogeneous syndrome has improved in recent years. Classification of adult and pediatric forms, based on histopathological data, into myopathic, neuropathic, or unclassified forms has been attempted, with each form occurring as a primary disorder or secondary to a nongastrointestinal disease (2, 3). In pediatrics, all studies have involved case reports or small series (4–11), except for one large survey reporting 87 cases for which no

Manuscript received July 23, 1998; accepted December 1, 1998.
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data about histopathology, therapeutic approach, or prognosis were available (12).

The aim of this study was to collect pediatric cases to determine clinical presentation and associated morbidity, to refine diagnostic and therapeutic approaches, and to describe outcome and prognosis of chronic intestinal pseusoobstruction in children.

MATERIALS AND METHODS

Each member of the French-Speaking Group of Pediatric Gastroenterology was sent a questionnaire. Physicians were asked to include patients who had been treated for CIP since 1980 in the study. The criteria for inclusion were the presence of recurrent episodes of intestinal obstruction with no mechanical obstruction or chronic constipation with small intestine distension visible on x-ray, with or without bilious vomiting, excluding Hirschsprung's disease. Premature infants were excluded.

Each questionnaire asked for the patient's initials, sex, date of birth, and the name of the referring center. Clinical presentation was assessed by description of family history, prenatal diagnosis, particularly postconceptional age at which the diagnosis was made, the presence of hydramnios, bladder distension, urinary tract distension, digestive loop distension, age at onset of symptoms, acute (defined as severe acute occlusive syndrome) or chronic mode of onset, the presenting symptoms, and any associated disease—particularly neurologic and myopathic disorders—disorders of other systems, or general disorders. The means and tools used for diagnosis were described in terms of a requirement for surgery at diagnosis, barium x-rays, urinary tract ultrasound scans, cystography, manometric recordings, and histopathologic data including whether full-thickness biopsies were taken, sites of biopsies, and histopathologic diagnosis. Treatment data included the type of nutritional support given with age of onset and duration of its use, the use of prokinetic agents and antibiotics to control bacterial overgrowth, the use of specific techniques to treat urinary tract complications and surgery. Follow-up and outcome were carefully noted in term of the age at which artificial feeding ceased. Gastrointestinal and nongastrointestinal complications were assessed, and if the patient died, the cause of death and age at death were recorded.

The histopathologic and manometric results are not reviewed in this study. Histopathologic diagnosis was classified on the basis of answers given in the questionnaires as neuropathy, myopathy, or unclassified. As different manometric techniques were used by the various centers, the results were not investigated further except for the presence or absence of rectoanal inhibitory reflex.

EpiInfo software was used for data analysis. Pair-wise comparisons of groups of patients were performed using the chi-square test.

RESULTS

In all, 109 completed questionnaires were collected from 17 centers. Four of the 109 were duplicates as the patients underwent follow-up in two different centers. This left 105 patients who fulfilled the criteria for inclusion in the study. Fifty-seven of the patients were boys and 48 were girls.

Family History

Five patients were from five different families with histories of CIP. In four of these families, only the siblings were affected. In two families, the diagnosis was of syndromic neuronal pseusoobstruction involving CIP, malrotation, short bowel, and pyloric hypertrophy. In two families, the affected patients presented with evident visceral myopathy. In one family, the father was affected, and there was no clear histopathologic diagnosis. Two other families were consanguinous.

Prenatal Diagnosis

There were prenatal signs of hollow viscera disease in 18 patients (17%). The signs were observed between 21 and 35 weeks after conception. The most frequent sign was megacystis (16 cases), followed by polyhydramnios (5 cases), and dilatation of upper urinary tract (5 cases). Dilatated intestinal loops were observed in only one case.

Clinical Presentation

Eighty patients were less than 12 months old at onset. Of these, the disease began at birth for 37 patients and during the first year of life for 43. The disease began before the end of the first month of life in 67% of cases.

Acute onset was reported in 64 cases; 53 patients were less than 1 year old, including 33 newborns. Thus, acute onset occurred in 89% of the newborns. The most frequent signs were abdominal distension (98%), vomiting (91%; bilious in 80%), constipation (77%), and failure to thrive (62%). Abdominal pain was reported in 58%, septicemia of digestive origin in 34% (32 patients), and diarrhea in 31% (29 patients). Dysphagia was rare (9 cases). Megacystis occurred in 31 patients and was noted in myopathies (7 cases), neuropathies (11 cases), and unclassified forms (13 cases).

The three main characteristic symptoms in patients with progressive symptoms were constipation, abdominal distension, and vomiting. Failure to thrive was reported in 72% of patients with progressive symptoms. There were other diseases present in 35 cases (Table 1).

TABLE 1. DISORDERS ASSOCIATED WITH CHRONIC INTESTINAL PSEUDOOBSTRUCTION

Disorders	N
Neurological ($N = 14$)	
Microcephaly	1
Areflexia	2
Perceptive deafness	1
Anoxia	2
Ependymoma, astrocytoma	2
Vermis hypoplasia	1
Callosum corpus agenesis	2
FG syndrome	1
Halerworden Spatz syndrome	1
MELAS*	1
Muscular ($N = 3$)	
Duchenne muscular dystrophy	1
Myotonic dystrophy	1
MELAS*	1
Cardiac ($N = 4$)	
Ventricular septal defect	1
Patent ductus arteriosus	1
Cardiomyopathy	1
Atrial septal defect	1
Uropathies, nephropathy (except megacystis) ($N = 6$)	
Hydronephrosis	2
Pyelic bifidity	1
Microcystis	1
Renal malrotation	1
IgA nephropathy	1
Infection ($N = 2$)	
CMV	2
Other ($N = 4$)	
Atopy	2
Interstitial desquamative Pneumopathy	1
Münchausen-by-proxy	1

* MELAS: mitochondrial myopathy, encephalopathy, lactic acidosis, strokelike episodes.

Diagnosis

Laparotomy. Surgery was used as a first-step diagnostic tool in 52 cases. Of these, 42 presented with acute onset, 43 were less than 1 year old, and 22 were newborns. Three patients underwent two laparotomies and one three laparotomies to diagnose their disease. Venting enterostomy was performed during the laparotomy in 31 cases. There were 19 ileostomies, 11 colostomies, and 1 jejunostomy. Only 9 patients underwent a single laparotomy with no other intervention during their disease.

Radiology. Barium opacification was used for all but two patients. Upper series were performed in 103 patients. Bowel dilatation was the most frequent sign, present in 62% of cases. Malrotation was seen in 9 cases, in all of which the age at onset was less than 2 months; 6 were neonates. Barium enema was performed in 98 patients. Colonic dilatation was observed in 58% of cases and microcolon in 10 cases. Barium enema results were normal in 17 patients. Eighty-two percent of patients had dilatation of at least one region of the gastrointestinal tract. Sixteen of the 18 patients with no dilatation were newborns or younger than 2 months.

Manometric Studies. Manometric studies were performed in 82 (79.8%) patients. Rectoanal manometry (72 patients) and esophageal manometry (71 patients) were the most used methods. Antroduodenal manometry was performed in 54 cases and colonic manometry in 15 cases. Rectoanal inhibitory reflex was detected in all but four cases.

Histopathology. At least one full-thickness biopsy from the digestive tract was studied for 99 patients. Rectal specimens were the most frequently available. The diagnosis recorded was visceral neuropathy in 58 cases and visceral myopathy in 17 cases. The diagnosis was uncertain or the biopsy considered normal in 24 cases.

Urinary Tract Involvement. Renal ultrasound scans were performed in 96 cases. Megacystis was found in 31 patients. There was dilatation of the upper urinary tract in 21 cases (22.6% of all patients, 68% of the patients with megacystis). Cystography was performed in 49 patients, and there was vesicorenal reflux in 13 cases.

Treatment

Nutritional Support. Eighty-seven children (83%) required nutritional support. Forty-four patients (42%) were given total parenteral nutrition (TPN), 35 (33%) were fed both parenterally and enterally by constant rate enteral nutrition (CREN), and 8 were fed enterally by CREN only.

Prokinetics and Antibiotics. Prokinetic drugs were used in 92% of the patients. Cisapride, bethanechol, erythromycin, and cerulein were the most commonly used. Bacterial overgrowth was treated and prevented with nonabsorbable antibiotics in 37% of the patients.

Surgery. Seventy-one patients underwent surgery as a treatment during their illness. There were 217 surgical procedures, a mean of 3 per patient (range: 1–14 interventions). Ostomy was the most performed procedure: 53 ileostomies, 17 colostomies, 3 jejunostomies, and 1 duodenostomy. These procedures were carried out during the first operation (52 cases) or the second operation (6 cases) in 58 of the 71 patients who underwent surgery. Twenty-two of these patients had the stoma closed after a mean of 39 months (range: 1–101 months) during the second operation (12 cases), the third operation (7 cases), or the fourth, fifth, or sixth operation (1 case each). The closure was

Table 2. Comparison Between Patients Requiring and No Longer Requiring Artificial Feeding

	Able to tolerate oral feeding (N = 38)	*Dependent on artificial feeding (N = 40)*	P
Prenatal signs	6	11	NS
Onset <1 year old	30	36	NS
Onset at birth	10	21	<0.05*
Acute onset	21	31	<0.05*
Megacystis	4	11	<0.05*
Associated disease	12	13	NS
Laparotomy at diagnosis	20	23	NS
Surgery	24	35	<0.05*

* Chi-square test.

performed following a Duhamel procedure in 11 cases, a Swenson procedure in 1 patient and simple closure in 10 cases. Gastrostomy was performed in 20 patients. Cystostomy was required in 6 cases. Ten patients had sequential cystic drainage, and 15 were treated with antiseptics. One boy, with familial visceral myopathy, underwent bowel transplantation at the age of 14 years after a total enterectomy for intractable occlusive syndrome.

Outcome

Follow-up continued in 89 patients for between 3 months and 16 years (mean 85 months) and 78 of them were fed nonorally during the course of their disease. Thirty-six ceased to require special forms of nutritional support after 1–144 months, at an age of 4 months to 20 years. In these 36 patients, parenteral nutrition was used in 33 for a mean of 18 months (range: 1–144 months) and enteral feeding in 25 for a mean of 13 months (range: 1–72 months).

Forty-two patients were still fed by parenteral (39 patients) or enteral nutrition (3 patients) at the time of the study. These 42 patients were given artificial feeding for between 2 months and 16 years. Eleven patients were given TPN for more than 10 years.

Twenty-four of the 58 patients who received ostomies were able to be orally fed and 34 are still dependent on artificial means of feeding. Twenty of the patients no longer requiring artificial feeding had their stoma closed.

Comparison between the two groups of patients, those requiring and those no longer requiring artificial feeding, showed significant differences in terms of neonatal onset, urinary tract involvement, acute onset, and requirement for surgery, all features that are more frequent in cases with a poor prognosis (Table 2).

Complications

Apart from cholelithiasis (6 cases), the most frequent were hematemesis (5 cases), diversion colitis (4 cases), and stoma prolapse (4 cases). Hypersecretion and excessive hydroelectrolytic losses were also reported in 3 patients. Two patients presented with medullary thyroid carcinoma associated with type IIb multiple endocrine neoplasia with neurogangliomatosis. Recurrent pancreatitis, gastric bezoar, gastric perforation, and gastric volvulus on gastrostomy were also reported in 4 different patients.

Mortality

Eleven patients died. The age at death was between 1 month and 14 years 7 months. The disease began when the patients were less than 1 year old in 9 of these patients. The cause of death was TPN-associated complications in 5 patients (hepatic failure in 3, infection of central line in 2). Septic shock of gastrointestinal origin was reported in 4 patients. Medullary thyroid carcinoma in a 14-year-old child with ganglioneuromatosis and sudden unexplained death at 2 years accounted for the 2 remaining cases.

DISCUSSION

Chronic intestinal pseudoobstruction is a rare disorder in children, and data are required to characterize clinical presentation, outcome, and prognosis. Studies in both children and adults, due to the rarity of the syndrome, tend to include only small numbers of cases or are focused on highly specific topics. We report here a study of 105 children diagnosed and treated for CIP in 17 centers.

Diagnosis of CIP has been controversial and has recently been defined in children as a clinical syndrome "characterized by repetitive episodes or continuous symptoms and signs of bowel obstruction, including radiographic documentation of dilatated bowel (except in a few congenital cases), in the absence of a fixed lumen occluding lesion" (13). In this study, all 105 patients included met these criteria and were therefore clearly suffering from intestinal pseudoobstruction. Indeed, CIP is diagnosed clinically after lumen obstruction has been ruled out, and histopathology and manometry are not formally required for diagnostic, but are means to characterize, classify and establish the prognosis (14).

The sex ratio shows that more boys than girls were affected in our sample. This is consistent with the survey of Vargas et al (12) but not with other studies (5–11). However, more newborn girls were affected

than newborn boys (23 girls *vs* 14 boys had the disease at birth), consistent with the previous reports (11). This suggests there may be a different neonatal or antenatal form of CIP.

Almost all patients presented with a dilated digestive segment on x-ray with opacification. The children with no bowel dilatation were very young, suggesting that dilatation could not occur at that stage or that the digestive tract was not functional. Malrotation was frequent, especially in neonates, and was reported in cases of myopathy, neuropathy, and familial syndrome with associated CIP, malrotation, and pyloric stenosis. Other authors have also noted a high prevalence of malrotation, especially in neonates (10–12).

Diagnosis was made after laparotomy, especially in cases of neonatal acute onset, to rule out lumen obstruction. Surgery may cause adhesions, so interpretation of subsequent obstructive episodes is difficult. This is reflected in the small number of patients who underwent a single laparotomy during their illness, suggesting that others underwent further surgery because occlusive episodes recurred, raising the possibility of adhesions. Thus, unnecessary surgery should be avoided, the typical case being a neonate with an antenatal diagnosis of megacystis without clear evidence of bowel obstruction on x-rays with opacifications. Indeed, surgery at diagnosis was avoided in more than 50% of our patients. However, if x-rays, manometry, or the clinical course of the illness suggest an organic obstructive lesion, laparotomy should be done to make formal exclusion possible. Almost all other pediatric reports note the high prevalence of surgery at diagnosis (4–12).

CMV infection was reported in two cases with infection of the ganglion cells, and case reports of these patients have already been published (15, 16). One of our cases was associated with a Münchausen-by-proxy syndrome (17). Except for these three patients, no case in this survey was secondary to a general disorder, which is usually the case in adults (18). This is a clear difference from adult CIP and suggests that pediatric CIP may be regarded as a specific entity usually related to primary anomalies of smooth muscle or the enteric nervous system. However, CIP is not always a primary disorder in children, and specific treatment can be given for curable infectious (18), inflammatory (19), or toxic (13) forms of CIP, for which there should therefore be systematic screening.

Five families were affected by familial forms of the illness. In two cases, the diagnosis of syndromic neuronal pseudoobstruction involving CIP, malrotation, short bowel, and pyloric hypertrophy was clearly established. This syndrome, first described by Royer et al (20), is thought to be transmitted with a recessive autosomal pattern of inheritance, but has been shown to be linked to Xq28 too (21). Familial visceral myopathies have also been reported in recessive (22, 23) and dominant forms (24, 25), some forms associated with ophthalmoplegia suggesting a mitochondrial myopathy. Our two patients with familial visceral myopathy seemed to have a recessive autosomal disease.

As reported for three patients, smooth muscle involvement has been reported in muscular diseases such as Duchenne muscular dystrophy (26), myotonic dystrophy (27), and mitochondrial myopathies (28). CIP is a rare feature but it is well characterized and should be treated as a primary visceral myopathy related to the striated and smooth muscle degenerative process (29).

Treatment methods and outcomes of CIP in children have not been reported for a large survey. Two thirds of patients required parenteral nutrition, indicating that most of our patients had severely impaired digestive motility, as suggested in studies showing that tolerance of oral feeding decreases as motility becomes more disturbed (10, 30, 31). Prokinetic drugs were tried in almost all patients with no clear efficacy (32). Treatment and prevention of bacterial overgrowth must be carefully managed in CIP because use of antibiotics leads to the development of resistant bacterial strains. Surgery was required as a treatment in 71 patients with a mean of three procedures per patient. This is similar to the situation noted by Krishnamurthy et al (9), with 67 operations in 22 patients. The most frequent procedure was digestive decompression to bypass the functional obstruction. Twenty-four of the 58 patients treated in this way clearly improved because it became possible to stop artificial feeding. This suggests that surgery was ineffective in more than 50% of these patients in terms of dependence on artificial feeding methods. However, these patients probably had less bacterial overgrowth, translocation, chronic distension, and abdominal pain as a result of surgical treatment. Surgery was also used for treatment in other small series, with variable results, ineffective in some (9, 12), and making it possible to stop artificial feeding for 4 of 10 patients in other (10). Small bowel transplantation was proposed in one boy as a last resort. Transplantation in CIP patients is complicated by specific problems such as gastric emptying disorders or achalasia (33).

Complications do occur, and some, such as stoma prolapse, recurrent pancreatitis, diversion colitis, and

excessive hydroelectrolytic losses have been reported in CIP patients (34–36) and should be considered as specific to CIP. Gastric perforation and gastric volvulus have never been reported with CIP.

The outcome was poor in the majority of our cases, despite the large number of children (46%) no longer requiring artificial feeding. Other reports do not give details of outcomes, except Fell et al (10) who reported a series of 14 cases with 5 enterally fed children, 4 TPN-dependent for 1–10 years, and 5 patients who died (10). Neonatal onset, urinary tract involvement, acute onset, and requirement for surgery indicate a poor prognosis. Indeed, neonatal forms of CIP have a high mortality rate and in the 10 living children reviewed by Granata and Puri (11), only one could tolerate enteral feeding. Urinary tract involvement reflects that this is a diffuse hollow viscera disease, suggesting that the entire digestive tract is involved in these cases, unlike the localized forms. The requirement for surgery may reflect the gravity of the disease.

Mortality was lower in this study than in other pediatric reports. Causes of death were directly related to CIP (septic shock of digestive origin, medullary thyroid carcinoma) or to parenteral nutrition (hepatic failure, infection of central line). Cardiac arrest has been reported in two previous cases of CIP and in one patient reported here (37).

In conclusion, we report a large survey of 105 pediatric cases of CIP. This rare syndrome has a specific pediatric form, which has been clearly described in terms of clinical presentation, management, and prognosis. Further studies should be performed to investigate the histopathological lesions of these patients.

ACKNOWLEDGMENTS

We would like to thank all members and nonmembers of the French-Speaking Group of Pediatric Gastroenterology who responded to the questionnaires.

REFERENCES

1. Schuffler M, Rohrman CA, Chaffe MD, et al: Chronic intestinal pseudo-obstruction: A report of 27 cases and review of the literature. Medecine 60:173–196, 1981
2. Krishnamurthy S, Schuffler MD: Pathology of neuromuscular disorders of the small intestine and colon. Gastroenterology 93:610–639, 1987
3. Navarro J, Sonsino E, Boige N, Nabarra B, Ferkadji L, Mashako LMN, Cezard JP: Visceral neuropathies responsible for chronic pseudo-obstruction syndrome in pediatric practice: Analysis of 26 cases. J Pediatr Gastroenterol Nutr 11:179–195, 1990
4. Byrne WJ, Cipel L, Euler AR, et al: Chronic idiopathic intestinal pseudoobstruction syndrome in children: Clinical characteristics and prognosis. J Pediatr 90:585–589, 1977
5. Anuras S, Mitros FA, Soper RT, et al: Chronic intestinal pseudo-obstruction in young children. Gastroenterology 91:62–70, 1986
6. Schuffler M, Pagon RA, Schwartz R, Bill AH: Visceral myopathy of the gastrointestinal and genitourinary tracts in infants. Gastroenterology 94:892–898, 1988
7. Nonaka M, Goulet O, Ahran P, Fekete C, Ricour C, Nezelof C: Primary intestinal myopathy, a cause of Intestinal pseudo-obstruction syndrome. Pediatr Pathol 9:409–424, 1989
8. Glassman M, Spivak W, Mininberg D, Marada J: Chronic intestinal pseudo-obstruction: A commonly misdiagnosed disease in infants and children. Pediatrics 83:603–608, 1989
9. Krishnamurthy S, Heng Y, Schuffler M: Chronic intestinal pseudo-obstruction in infants and children caused by diverse abnormalities of myenteric plexus. Gastroenterology 104:1398–1408, 1993
10. Fell JME, Smith VV, Milla P: Infantile chronic idiopathic intestinal pseudo-obstruction: The role of small intestinal manometry as a diagnostic tool and prognostic indicator. Gut 39:306–311, 1996
11. Granata C, Puri P: Megacystis-microcolon-intestinal hypoperistalsis syndrome. J Pediatr Gastroenterol Nutr 25:12–19, 1997
12. 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 7:323–332, 1988
13. Rudolph CD, Hyman PE, Altschuler SM, et al: Diagnosis and treatment of Chronic intestinal pseudo-obstruction in children: Report of a consensus workshop. J Pediatr Gastroenterol Nutr 24:102–112, 1997
14. Schuffler MD: Chronic intestinal pseudoobstruction: Progress and problems. J Pediatr Gastroenterol Nutr 10:157–163, 1990
15. Sonsino E, Mouy R, Foucaud P, et al: Intestinal pseudo-obstruction related to cytomegalovirus infection of myenteric plexus. N Engl J Med 311:196–197, 1984
16. Ategbo S, Turck D, Gottrand F, et al: Chronic intestinal pseudoobstruction associated with cytomegalovirus in an infant. J Pediatr Gastroenterol Nutr 23:457–460, 1996
17. Ginies JL, Goulet O, Champion G, et al: Munchausen's syndrome by proxy and chronic intestinal pseudo-obstruction. Arch Fr Pediatr 46:267–269, 1989
18. Christensen J, Orvar K: Chronic pseudo-obstruction. *In* An Illustrated Guide to Gastrointestinal Motility. D Kumar, D Wingate (eds). London, Churchill Livingstone, 1993, pp 567–582
19. Ginies JL, Francois H, Joseph MG, Champion G, Coupris S, Limal JM: A curable cause of intestinal pseudoobstruction in children: Idiopathic myositis of the small intestine. J Pediatr Gastroenterol Nutr 23:426–429, 1996
20. Royer P, Ricour C, Fekete C, et al: Le syndrome familial de grêle court avec malrotation intestinale et sténose hypertrophique du pylore chez le nourrisson. Arch Fr Pediatr 31:223–229, 1974
21. Auricchio A, Brancolini V, Casari G, et al: The locus for a novel syndromic form of neuronal intestinal pseudoobstruction maps to Xq28. Am J Hum Genet 58:743–748, 1996
22. Anuras S, Mitros FA, Nowak T, et al: A familial visceral myopathy with external ophthalmoplegia and autosomal recessive transmission. Gastroenterology 84:346–353, 1983

23. Jacob E, Ardichvili D, Rerissino A, Gottinies P, Hansens JF: A case of familial visceral myopathy with atrophy and fibrosis of the longitudinal muscle layer of the entire small bowel. Gastroenterology 77:745–750, 1979
24. Schuffler M, Pope CE: Studies of idiopathic intestinal pseudoobstruction. II. Hereditary hollow visceral myopathy family studies. Gastroenterology 73:339–344, 1977
25. Rodrigues CA, Shepperd NA, Lennard Jones JE, Hawley PR, Thompson HH: Familial visceral myopathy: A family with at least six involved members. Gut 30:1285–1292, 1989
26. Leon SH, Schuffler MD, Kettler M, Rohrmann CA: Chronic intestinal pseudoobstruction as a complication of Duchenne's muscular dystrophy. Gastroenterology 90:455–459, 1986
27. Bruinenberg JFM, Rieu PNMA, Gabreels FM, Tolboom J: Intestinal pseudoobstruction syndrome in a child with myotonic dystrophy. Acta Paediatr 85:121–123, 1996
28. Lowsky R, Davidson G, Wolman S, Jeejeebhoy KN, Hegele RA: Familial visceral myopathy associated with a mitochondrial myopathy. Gut 34:279–283, 1993
29. Miyatake M, Mike T, Yoshioka K, Uchino M, Usuku G: Possible systemic smooth muscle layer dysfunction due to a deficiency of dystrophin in Duchenne muscular dystrophy. J Neurol Sci 93:11–17, 1989
30. Boige N, Faure C, Gargill G, et al: Manometrical evaluation in visceral neuropathies in children. J Pediatr Gastroenterol Nutr 19:71–77, 1994
31. Cucchiara S, Annese V, Minella R, et al: Antroduodenojejunal manometry in the diagnosis of chronic intestinal pseudoobstruction in children. J Pediatr Gastroenterol Nutr 18:294–305, 1994
32. Camillieri M, Balm RK, Zinsmeister AR: Determinants of response to a prokinetic agent in neuropathic chronic intestinal motility disorder. Gastroenterology 106:916–923, 1994
33. Goulet O, Jan D, Brousse N, Revillon Y, Ricour C: Intestinal transplantation. J Pediatr Gastroenterol Nutr 25:1–11, 1997
34. Heitlinger LA, McClung HJ, Murray RD, Li BUK: Recurrent pancreatitis in three patients with chronic idiopathic intestinal pseudoobstruction. J Pediatr Gastroenterol Nutr 13:92–95, 1991
35. Ordein JJ, Di Lorenzo C, Flores A, Hyman PE: Diversion colitis in children with severe gastrointestinal motility disorders. Am J Gastroenterol 87:88–90, 1992
36. Rolston DDK, Hunt JB, Fairclough PD, et al: Jejunal water and sodium secretion occurs in chronic idiopathic intestinal pseudoobstruction. J Clin Gastroenterol 12:153–156, 1990
37. Anuras S, Mitros FA, Shirasi SS: Cardiac arrest in two children with non familial chronic intestinal pseudoobstruction on total parenteral nutrition. J Pediatr Gastroenterol Nutr 1:137–144, 1982