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Visceral Myopathy of Intestinal Pseudoobstruction

Sang-Yong Song, Je G. Chi[1], Jeong Kee Seo[2], Jung-Hwan Choi[2] and Kwi Won Park[3]

Department of Pathology, Pediatrics[2] and Pediatric Surgery[3], Seoul National University College of Medicine and Seoul National University Children's Hospital, Seoul 110-799, Korea

= Abstract = **Intestinal pseudoobstruction is a syndrome complex caused by a variety of disorders of various etiology. It can be classified pathologically as visceral myopathy and visceral neuropathy. The sporadic form of visceral myopathy is characterized histologically by vacuolar degeneration and fibrosis of smooth muscle but differs from the familial form only by the absence of other affected family members. We studied 6 cases with intestinal pseudoobstruction classified as sporadic visceral myopathy. They were four boys and two girls, and were two neonates, two infants and two children. The duration of symptoms ranged from two days to two years. Two babies were dead from pneumonia and sepsis. Others were alleviated after surgical resection of the bowel. Both small and large intestines were found affected in autopsy cases. Histopathologic features were vacuolar degeneration of muscularis propria, disproportionate hypoplasia of outer muscle layer, abnormal muscle direction of muscularis propria, submucosal and/or interstitial fibrosis and extra muscle layering. It is presumed that a variety of histopathologic features accounts for visceral myopathy of intestinal pseudoobstruction.**

Key Words: *Intestinal pseudoobstruction, Visceral myopathy, Megacystis microcolon intestinal hypoperistalsis syndrome, Childhood*

INTRODUCTION

Intestinal pseudoobstruction is a clinical syndrome characterized by symptoms and signs of intestinal obstruction without any concrete evidence for an actual lesion obstructing the intestinal lumen(Faulk *et al.* 1978). It can be divided into acute or transient form and chronic or recurrent form by the duration of the symptoms. The chronic form can further be classified into either primary (idiopathic) or secondary(associated with progressive systemic sclerosis, amyloidosis, myotonic dystrophy, or Chagas' disease) (Mitros *et al.* 1982; Morson *et al.* 1990). In general, so-called intestinal pseudoobstruction means primary(idiopathic) chronic intestinal pseudoobstruction. Chronic idiopathic intestinal pseudoobstruction(CIIP) has no underlying systemic disease and can be divided into familial and sporadic type. In addition, it can also be classified as smooth muscle disorders, neurological disorders, or no detectable abnormalities(Faulk *et al.* 1978; Mitros *et al.* 1982; Morson *et al.* 1990). Familial visceral myopathy is apparently the most common cause and the most thoroughly studied form of CIIP(Mitros *et al.* 1982). Although both

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[1]Author for correspondence : Tel. 02-760-2550
서울대학교 의과대학 병리학교실 및 어린이병원 병리과 : 송상용, 지제근
서울대학교 의과대학 소아과학교실 및 어린이병원 소아과 : 서정기, 최중환
서울대학교 의과대학 외과학교실 및 어린이병원 소아외과 : 박귀원

the familial and sporadic forms of visceral myopathy are characterized by vacuolar degeneration and fibrosis of the smooth muscle(Mitros *et al.* 1982; Schuffler *et al.* 1988), the histopathologic characteristics of sporadic visceral myopathies have not been fully elucidated. Based on our cases, we describe the histopathologic features of visceral myopathy of CIIP trying to expand the pathologic spectrum of the disease.

CASE REPORTS

Case 1

A three-day-old male neonate presented with abdominal distention and fever of 2 days duration. He was born by normal full-term spontaneous vaginal delivery to a 29-year-old mother without perinatal problems. Physical examination and radiologic study revealed dilated stomach and small intestine, intestinal malrotation and small calibered colon. Voiding cystourethrography demonstrated a marked vesical distention neither with vesico: ureteral reflux nor with urethral obstruction. Exploratory laparatomy showed distended jejunum and proximal ileum, and narrowed distal ileum with shortening of the entire small bowel length. There was no demonstrable mechanical obstruction. The blood culture done on admission grew Staphylococcus species, and the fever was controlled. However, intestinal activity was very poor, and neither gastrografin enema nor neostigmin were effective for meconium passage. He died on the 42nd day of life due to sepsis. At autopsy the body weighed 2,100 gm and showed marked emaciation with jaundice and abdominal distention. There was a markedly dilated stomach, malfixed and dilated small intestine, and small calibered colon with a

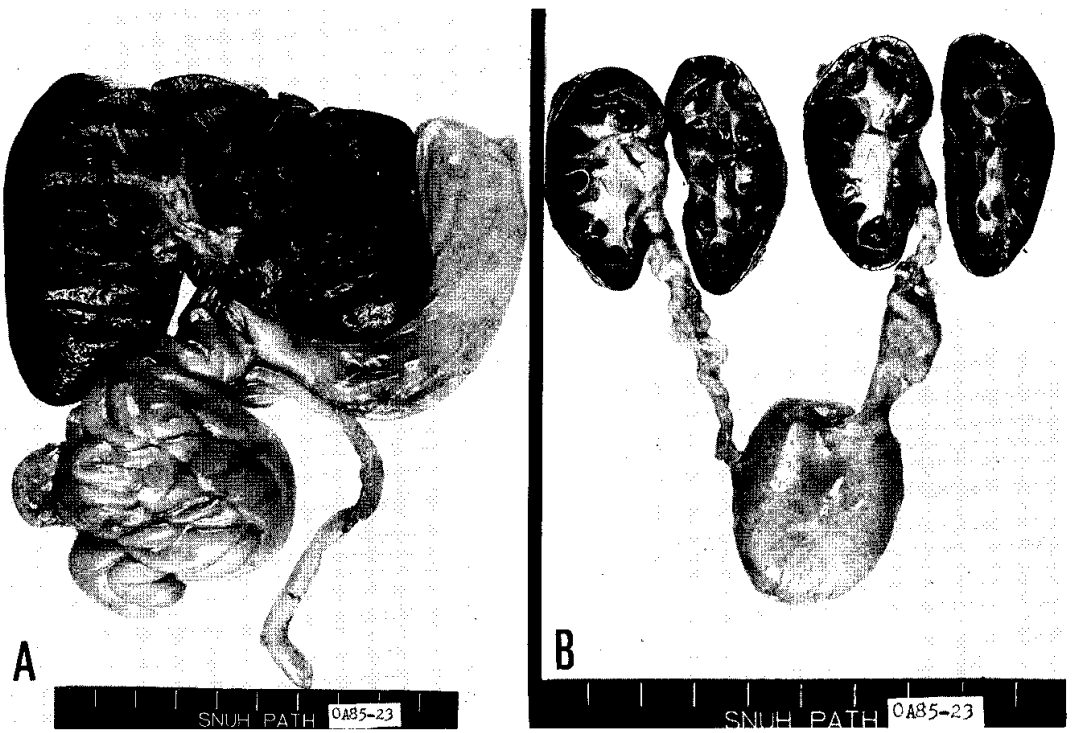

Fig. 1. Postmortem examination of case 1 shows (A) microcolon with intestinal malrotation and (B) megacystis with hydroureter.

mean diamter of 0. 4 cm(Fig. 1A). The rectum and anus were unremarkable. The urinary bladder was diffusely dilated into a round shape (Fig. 1B). The urethra was patent. Both kidneys weighed 17 gm together and showed a considerable hydropelvis. There was no evidence of obstruction in any portion of the urinary tract. Microscopically, sections from the small and large intestine showed mild vacuolar degeneration, disproportionate hypoplasia of the outer muscle layer and submucosal fibrosis. Ganglion cells in submucosa and muscularis propria were well preserved in the entire segment of the small and large intestine.

Case 2

A 97-day-old female infant presented with low birth weight, respiratory distress and abdominal distention since birth. She was delivered after 28 weeks of gestation by Caesarean section because of total placenta previa and premature rupture of the mother's membrane. She weighed 857 gm at birth and stained with meconium. Apgar scores were 6 at 1 minute and 7 at 5 minute, respectively. Occult blood test was positive in the stool. The symptoms waxed and waned, and ventricular dilatation and periventricular leukomalacia were noted in the brain sonogram. She died of sepsis.

At autopsy the body weighed 1,140 gm and showed marked emaciation with abdominal distention. On opening the peritoneal cavity, hepatosplenomegaly and ascites were noted but the urinary bladder and gastrointestinal tract were unremarkable grossly. The lungs showed diffuse consolidation and hemorrhage. Microscopically, sections from the small and large intestine showed vacuolar degeneration, and irregular thinning and thickening of the inner muscle layer(Fig. 2).

Characteristically, the direction of the inner muscle layer was parallel with the longitudinal axis of the intestine. The outer muscle layer also showed abnormal direction reminiscent of the inner circular muscle and showed segmental disproportionate hypoplasia. The submucosa showed fibrosis and well formed ganglion cells.

Case 3

A 4-year-old boy presented with abdominal distention and vomiting which he had suffered since 6 months of age. He underwent Duhamel's operation for congenital megacolon.

Fig. 2. Photomicrograph of case 2 shows vacuolar degeneration of the inner muscle layer and varying degrees of interstitial fibrosis. (Inset : high power photomicrograph of vacuolar degeneation of muscularis propria)

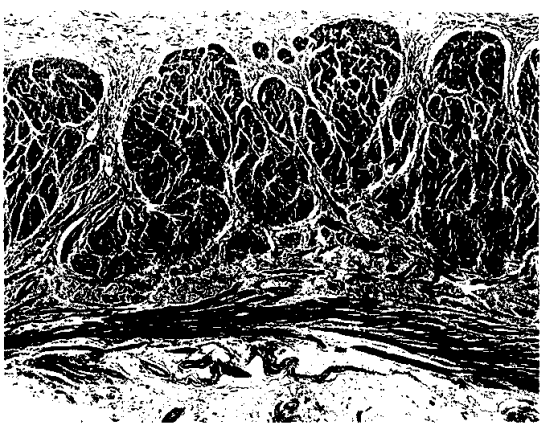

Fig. 3. Photomicrograph of case 3 shows severe interstitial fibrosis, irregularly arranged inner muscle layer and disproportionate hypoplasia of the outer muscle layer.

However, he was not confirmed histopathologically and did not show any improvement. In spite of adhesiolysis, gastrostomy, ileostomy and total parenteral nutrition, the symptoms aggravated. Therefore, he underwent subtotal gastrectomy and subtotal colectomy. Postoperative course was satisfactory.

The resected stomach and subtotal ileocolectomy specimen measured 12 cm and 48 cm in length, respectively. The mucosal folds of the intestine were flattened and the wall was irregularly thinned. Multiple sections showed segmental hyperplasia of the inner muscle layer, interstitial fibrosis and mild vacuolar degeneration of the muscularis propria(Fig. 3). The outer muscle layer was hypoplastic segmentally. Ganglion cells were prominent in both submucosal and myenteric plexuses. The submucosa showed fibrosis. Mononuclear cell infiltration was noted in both mucosa and subserosa.

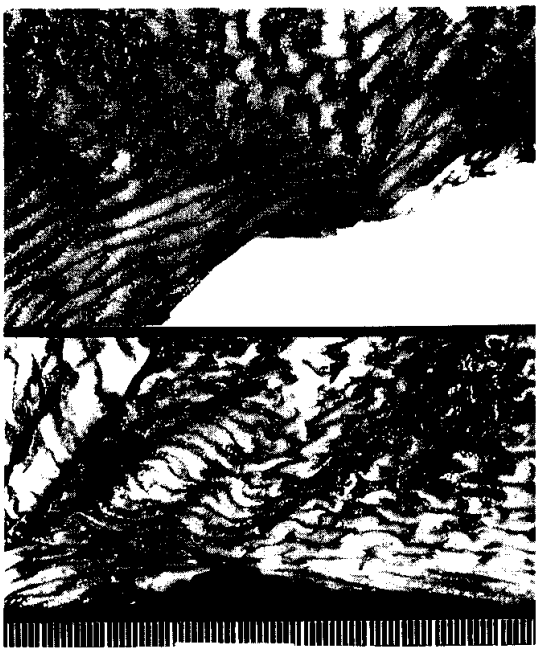

Fig. 4. Macroscopically, the mucosal surface of case 4 shows a haphazard arrangement of the mucosal folds.

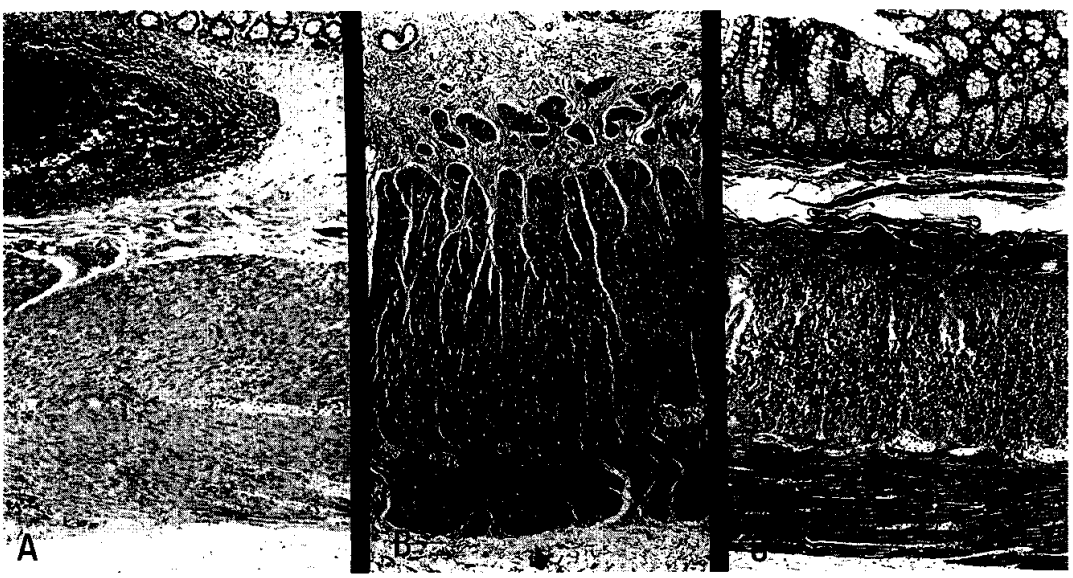

Fig. 5. Photomicrograph of case 4 demonstrates the varied histopathology of myopathy in segmental fashion including (A) abnormal direction of both inner and outer muscle layers, (B) incomplete additional muscle layering of irregularly proliferating smooth muscle bundles with abnormal direction, and (C) complete additional muscle layering with normal direction(Masson's trichrome).

Case 4

A two-year-old boy presented with constipation of one year's duration. At birth, he was unremarkable except for an imperforate anus, for which Pena operation was done at the age of one year. Though upper gastrointestinal series revealed malrotation of the small intestine with delayed barium passage, there was no demonstrable obstruction site. Radiolo- gically, congenital megacolon was suspected due to a marked dilation of the sigmoid colon. He underwent subtotal colectomy.

The resected colon measured 30 cm in length and showed irregular widening of the circumference up to 15 cm. Mucosal folds showed irregular direction with predominant longitudinal component(Fig. 4). The wall was irregularly thinned. Microscopically, a segment of inner muscle layer showed abnormal direction that was parallel to(Fig. 5A) or oblique to(Fig. 5B) the longitudinal axis of the intestine. Additional layering of irregularly proliferating muscle bundles was found(Fig. 5B and 5C). Focal disruption and vacuolar degeneration were also seen in the inner muscle layer. Ganglion cells were well present in the entire colonic segment.

Case 5

A 4-day-old female neonate presented with abdominal distention and vomiting which she had suffered from one day of age. The delivery was uneventful and the apgar score was 9. She was small for her age (birth weight 2.6 kg, gestational age 36 weeks). Radiologic examination revealed small-calibered distal ileum and colon with delayed barium passage which was suggestive of ileal stenosis or total colonic aganglionosis. The resected small intestine measured 28 cm in length and showed segmental thinning and narrowing. The narrowed portion measured 2 cm in diameter.

Mucosa was focally eroded. Microscopically, the inner muscle layer showed mild vacuolar change. The outer muscle layer showed disproportionate hypoplasia and segmental attenuation(Fig. 6).

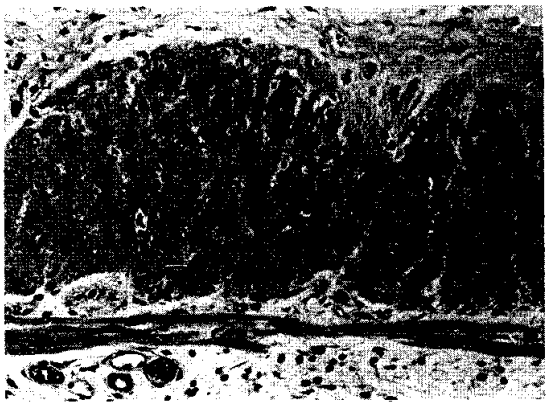

Fig. 6. Photiomcrograph of case 5 shows disproportionate hypoplasia and segmental attenuation of the outer muscle layer.

Ganglion cells were well present. Mononuclear cell infiltration was seen in the subserosa. Mild fibrosis was noted in the submucosa and subserosa.

Case 6

A two-month-old male infant presented with chronic diarrhea which he had had since birth. The delivery was uneventful except for mild meconium staining(birth weight 3.5 kg, gestational age 39 weeks). Antibiotic therapy was given because of continuous fever. However, he was not confirmed microbiologically and did not show any improvement. Radiologic impression was gastric volvulus. An open biopsy was done. The biopsy specimen consisted of 4 parts including the jejunum, ileum, appendix and colon. Histopathologically, all of them showed the same features. Muscularis propria exhibited vacuolar degeneration and submucosa showed fibrosis. Ganglion cells were well present in the entire segment.

We have summarized the clinical and pathologic features of these cases(Table 1).

DISCUSSION

Since the first description(Ogilvie 1948), many investigators(Swenson and Fisher 1955; Dudley *et al.* 1958; Davidson and Bauer 1958;

Table 1. Clinico-pathologic summary of the cases

Case	1	2	3	4	5	6
Clinical summary						
Sex/Age	M/3 ds	M/3 mos	M/4 yrs	M/2 yrs	M/4 ds	M/2 mos
Location	SI, LI	SI, LI	LI	LI	SI	SI, LI
Duration	2 ds	3 mos	2 yrs	1 yr	3 ds	2 mos
Fate	Expired	Expired	Alive	Alive	Alive	Alive
Specimen	Autopsy	Autopsy	Resection	Resection	Resection	Biopsy
Pathologic features						
Submucosa						
—Fibrosis	+	+	++	+	±	++
Muscularis propria						
—AML	—	—	—	+	—	—
—VD	±	+	±	+	±	+
—AD	—	+	—	+	—	—
—IF	±	+	+	+	±	—
—DH or SDH	±	+	+	+	+	—

ds : days, mos : months, yr(s) : year(S), SI : small intestine, LI : large intestine, AML : abnormal muscle layering, VD : vacuolar degeneration, AD : abnormal direction, IF : interstitial fibrosis, DH : disproportionate hypoplasia, SDH : segmental DH. Case 1 was previausly reported.

Table 2. Summary of related terminology of intestinal pseudoobstruction

year	Authors	Terminology
1948	Ogilvie	Large intestine colic due to sympathetic deprivation. A new clinical syndrome
1955	Swenson & Fisher	Megacolon and megaureter
1958	Dudley et al.	Intestinal pseudoobstruction
	Davidson & Bauer	Achalasia of distal rectal segment
1963	Siber & Girdany	Functional intestinal pseudoobstruction
1964	Bentley	Hypoganglionosis
1965	Ehrenpreis	Pseudo—Hirschsprung's disease
1966	Nixon	Chronic adynamic ileus
1970	Maldonado et al.	Chronic idiopathic intestinal pseudoobstruction
1975	Kapila et al.	Chronic adynamic bowel
1976	Berdon et al.	Megacystis microcolon intestinal hypoperistalsis syndrome
1977	Puri et al.	Adynamic bowel syndrome
	Schuffler et al.	Hereditary hollow visceral myopathy
	Puri et al.	Neuronal colonic dysplasia
1978	Faulk et al.	Familial visceral myopathy
1980	Schuffler et al.	Sporadic visceral myopathy
1984	Kirtane et al.	Dysganglionosis

Sieber and Girdany 1963; Bentley 1964; Ehrenpreis 1965; Nixon 1966; Maldonado *et al.* 1970; Kapila *et al.* 1975; Berdon *et al.* 1976; Puri *et al.* 1977; Schuffler *et al.* 1977; Puri *et al.* 1977; Faulk *et al.* 1978; Schuffler and Deitch 1980; Kirtane *et al.* 1984) have described the clinical syndrome characterized by symptoms and signs of intestinal obstruction without actual obstructing lesion under various names (Table 2). Most of these names are confusing and are not

pathologically oriented, indicating that it has long been a "clinical" entity without full elucidation of its histopathologic features. Morphologic examination of the gastrointestinal tract in such a lesion has largely been neglected(Mitros *et al.* 1982), until the neurologic and the myologic abnormalities were recently described (Table 3). Neurologic abnormality was focused

Table 3. Pathologic features according to the type of intestinal pseudoobstruction

A. Neuropathic type
  1) Abnormal neuronal structures and reduced number of ganglion and nerve bundles
  2) Degeneration of myenteric plexus
  3) Intranuclear inclusion and reduced number of neurons
  4) Reduced argyrophilic neurons, axonal swelling and fragmentation, and schwann cell scar
  5) Shrunken or pyknotic neurons-dysganglionosis

B. Myopathic type
  1) Hypertrophy of one or both muscle layers
  2) Atrophy and fibrosis of one or both muscle layers with/without increased nerves
  3) Vacuolar degeneration and fibrosis of smooth muscle
  4) Muscular hypertrophy, excessive oblique muscle layer with prominent and abnormal shaped myenteric plexus but normal ganglion cells

mainly on the Auerbach's plexus. Abnormal neuronal structure and reduced number of ganglion cells and nerve bundles(Erskine 1963; Smith 1968), degeneration of myenteric plexus (Dyer *et al.* 1969; Cockel *et al.* 1973), and intranuclear inclusion in nerve cells(Schuffler *et al.* 1978) were described. With the use of Smith's silver stain, Schuffler and Jonak showed patchy loss of nerve tracts with replacement by Schwann cells, degeneration and decreased numbers of both argyrophilic and argyrophobic neurons, fragmentation and drop-out of many axons, and increased thickness and disorganized spatial arrangement of other axons(Schuffler and Jonak 1982). They also focused a significantly decreased number of neurons compared with controls. In 1984, Kirtane *et al.* proposed a new terminology, "dysganglionosis", based on their observation of shrunken or pyknotic neurons in the myenteric plexus. Myologic abnormalities have been more variable and more obvious than the neurologic abnormalities because they could be found by routine histologic examination. Earlier reports described the hypertrophy of one or both smooth muscle layers(Naish *et al.* 1960; McCllend *et al.* 1962). However, the common histologic features were atrophy and fibrosis of one or both muscle layer with or without increased nerves(Murley 1959; Schuffler *et al.* 1977; Jacobs *et al.* 1979; Young *et al.* 1981; Puri *et al.* 1983; Bagwell *et al.* 1984), and vacuolar degeneration with fibrosis of the smooth muscle(Mitros *et al.* 1982; Bagwell *et al.* 1984). In 1988, Yamagiwa *et al.* reported a case of intestinal pseudoobstruction with massive muscular hypertrophy and excessive oblique muscle layer forming three layering of smooth muscles. The myenteric plexuses were prominent and more irregularly shaped than normal, but myenteric and submucosal ganglion cells were normal. Also, some investigators observed flattening of mucosal villi in the small intestine (Paul *et al.* 1961).

Our cases are included in the "intestinal pseudoobstruction complex" because all of our cases showed symptoms and signs of intestinal obstruction without any evidence of organic obstruction. Although case 3 and case 5 were clinically diagnosed as aganglionosis, histopathologic examination revealed well formed ganglion cells and therefore was of no organic cause. Case 1 is a typical case of megacystis — microcolon - intestinal - hypoperistalsis syndrome. We believe that this syndrome should be included in the intestinal pseudoobstruction complex. Histologic examination of our cases showed ; (a) vacuolar degeneration of the muscularis propria, (b) disproportionate hypo-

Table 4. Proposed pathologic classification of intestinal pseudoobstruction

1. Neuropathic type
   a. Aganglionosis : Achalasia, Hirschsprung's disease
   b. Dysganglionosis : CIIP
   c. Hypoganglionosis : CIIP
2. Myopathic type
   a. Hypertrophic : thickenning
   b. Atrophic : thinning, disproportionate hypoplasia.
   c. Dysmorphic : abnormal direction, in situ degenerationd.
   d. Hyperplastic : excessive or additional muscle proliferatione.
   e. Mixed
3. Combined neuropathic and myopathic type
4. Urinary counterpart
   a. Isolated form : Prune-belly syndrome
   b. Combined form : MMIHS
5. True idiopathic type

CIIP : Chronic idiopathic intestinal pseudoobostruction, MMIHS : Megacystis-microcolon-intestinal-hypoperistalsis syndrome

plasia of the outer muscle layer, (c) varying degrees of submucosal fibrosis, (d) abnormal muscle direction, (e) interstitial fibrosis of the muscularis propria, and (f) additional muscle layering. Though most of these features have been previously reported, abnormal muscle direction with additional muscle layering has not been described. Macroscopically, these cases showed abnormal and irregular direction of mucosal folds, suggesting that abnormal direction of the underlying muscle layer might be related to the direction of mucosal fold. We believe this lesion might be congenital rather than acquired, because the longitudinal mucosal folds are common findings of the fetal or neonatal colon. In comparison with previously reported cases, vacuolar degeneration in our cases was rather mild. Although the large intestine showed varying thickness of muscularis propria in the circumferential section, that of the small intestine is constant. Therefore, it is meaningful to use disproportionate hypoplasia in our case 2 and case 5. Furthermore, it can also be used in the colon because this change is segmental in our case 3 and case 4.

Three of our cases showed short duration of symptoms though histologic features were within CIIP. Therefore, CIIP may not be adequate to encompass all these features. We propose a classification of CIIP(Table 4). It can be classified as neuropathic type, myopathic type, combined neuropathic and myopathic type, urinary counterpart and true idiopathic type. Although some investigators regarded Hirschsprung's disease as secondary intestinal pseudoobstruction, we think Hirschsprung's disease belongs to the neuropathic type of intestinal pseudoobstruction because its pathology is present in the myenteric plexus without any pathology in other systems. Therefore Hirschsprung's disease can be classified as primary disease of the intestine. Since neuropathic change can be detected only by frozen section using Smith's silver stain(Schuffler and Jonak 1982), we were not able to evaluate our cases in this aspect. It seems necessary that a portion of an intestinal sample should be reserved for frozen section.

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