Chronic intestinal pseudo-obstruction: treatment and long term follow up of 44 patients

S Heneyke, V V Smith, L Spitz, P J Milla

Abstract

Aims—To document the long term course of chronic idiopathic intestinal pseudo-obstruction syndrome (CIIPS) in children with defined enteric neuromuscular disease, and the place and type of surgery used in their management; in addition, to identify prognostic factors.

Methods—Children with CIIPS were investigated and treated prospectively.

Results—Twenty four children presented congenitally, eight during the 1st year of life, and 10 later. Twenty two had myopathy and 16 neuropathy (11 familial). Malrotation was present in 16 patients, 10 had short small intestine, six had non-hypertrophic pyloric stenosis, and 16 had urinary tract involvement. Thirty two patients needed long term parenteral nutrition (TPN): for less than six months in 19 and for more than six months in 13, 10 of whom are TPN dependent; 14 are now enteral feeding. Prokinetic treatment improved six of 22. Intestinal decompression stomas were used in 36, colostomy relieved symptoms in five of 11, and ileostomy in 16 of 31. A poor outcome (death (14) or TPN dependence (10)) was seen with malrotation (13 of 16), short small bowel (eight of nine), urinary tract involvement (12 of 16), and myopathic histology (15 of 22).

Conclusions—In CIIPS drugs are not helpful but decompression stomas are. Outcome was poor in 24 of 44 children (15 muscle disorder, 10 nerve disease).

(Keywords: chronic intestinal pseudo-obstruction; decompression stomas; prognostic factors; total parenteral nutrition)

Chronic idiopathic intestinal pseudo-obstruction syndrome (CIIPS) is an intestinal motility disorder in which impaired intestinal motor activity causes recurrent symptoms of intestinal obstruction in the absence of mechanical occlusion. It is usually caused by disease of the enteric neuromusculature and may involve either segments of, or the entire, gastrointestinal tract. It is an uncommon disorder with a high morbidity and mortality. Until now, sufficient data were not available for the natural history of CIIPS to be apparent, and the place of surgery in its management is still not well defined.
Table 1  Clinical details of 44 patients with chronic intestinal pseudo-obstruction

<table>
<thead>
<tr>
<th>Patients</th>
<th>Weight centile Birth</th>
<th>Present</th>
<th>Age</th>
<th>Symptoms</th>
<th>Radiology</th>
<th>Histology</th>
<th>Surgery</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>1–6</td>
<td>3rd to 50th</td>
<td>–</td>
<td>Birth</td>
<td>Vomiting</td>
<td>Dilated whole gut</td>
<td>M</td>
<td>5</td>
<td>4 dead</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Intestinal obstruction</td>
<td>Pyloric obstruction</td>
<td>1</td>
<td>N</td>
<td>2 TPN</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Short small intestine</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>7–13</td>
<td>3rd to 25th</td>
<td>–</td>
<td>Antenatal</td>
<td>Vomiting</td>
<td>Malrotation</td>
<td>M</td>
<td>7</td>
<td>4 dead</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Failure to pass urine</td>
<td>Intestinal obstruction</td>
<td></td>
<td></td>
<td>1 enteral</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Dilated bowel</td>
<td></td>
<td></td>
<td>2 TPN</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Megacystis</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Megaureter</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>14–24</td>
<td>3rd to 50th</td>
<td>&lt; 3rd to 10th</td>
<td>Birth to 7 years</td>
<td>Constipation</td>
<td>3 malrotation</td>
<td>M</td>
<td>8</td>
<td>3 dead</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Obstruction</td>
<td>Megacystis</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Abdominal distension</td>
<td>Megaureter</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Dilated bowel</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Slow transit</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>25–30</td>
<td>10th to 75th</td>
<td>3rd to 25th</td>
<td>Birth to 2 years</td>
<td>Constipation</td>
<td>10 contracted colon</td>
<td>N</td>
<td>6</td>
<td>2 gastrostomy</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Dilated bowel</td>
<td>Abdominal distension</td>
<td></td>
<td></td>
<td>4 normal feeds</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>1 stricture</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>31–42</td>
<td>3rd to 50th</td>
<td>&lt; 3rd to 10th</td>
<td>Birth to 3 years</td>
<td>Constipation</td>
<td>3 malrotation</td>
<td>N</td>
<td>5</td>
<td>3 dead</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Obstruction</td>
<td>Abdominal distension</td>
<td></td>
<td></td>
<td>7 enteral</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Dilated bowel</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>1 contracted colon</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Slow small intestine</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>43</td>
<td>10th to 25th</td>
<td>&lt; 3rd to 10 years</td>
<td>2 years</td>
<td>Obstruction</td>
<td>Severe constipation</td>
<td>M</td>
<td>6</td>
<td>2 TPN</td>
</tr>
<tr>
<td>44</td>
<td>10th to 25th</td>
<td>&lt; 3rd to 10 years</td>
<td>10 years</td>
<td>Abdominal distension</td>
<td>Severe constipation</td>
<td>N</td>
<td>4</td>
<td>TPN</td>
</tr>
</tbody>
</table>

Present, presentation; M, myopathy; N, neuropathy; ?, unknown; TPN, long term total parenteral nutrition.

Small intestinal manometry

We assessed motor activity in the duodenum manometrically in 30 patients investigated since 1981 after an overnight 15 hour fast as described previously.11 We recorded fasting motor activity for three interdigestive cycles, where this was present, or for four hours, whichever was longer. The manometric patterns were analysed by means of visual planimetry by two blinded independent observers.11 Because the infants were vomiting and had a severe intolerance to feeds, postprandial activity was not studied.

STATISTICS

Fisher's exact probability test was applied to determine whether there were any significant prognostic factors.

Results

The criteria for the diagnosis of CIIPS were fulfilled by 44 patients (25 boys, 19 girls). Histopathology was available in all and motility, contrast, and marker transit studies in most. Thirty of the 44 patients had survived at the time of our study and had a mean age of 12 years (range, 3 months to 26 years). Details of all 44 patients are summarised in table 1.

ONSET OF SYMPTOMS AND CLINICAL PRESENTATION

Twenty four children developed obstructive symptoms during the 1st month of life: 18 were symptomatic at birth and seven were evident on ultrasound scanning antenatally. A further eight developed symptoms during the 1st year of life and 10 before the 3rd year of life. Two children did not develop constipation or obstructive symptoms until the age of 6 and 10 years, respectively. Seven had evidence of active disease in utero with an obstructed urinary system on antenatal ultrasound.

ULTRASOUND AND RADIOLOGICAL INVESTIGATION

All patients had a whole gut transit time of > 96 hours. Key radiological features are shown in table 1. Ten neonates presented with abdominal distension and bilious vomiting and an abdominal x ray suggestive of distal bowel obstruction. Seven neonates presented with the diagnosis of an obstructed urinary system on antenatal ultrasound scanning and failure to pass urine. Symptoms of intestinal obstruction developed during the 1st month of life in six and by the 4th month of life in two other infants. Five neonates, presenting with bilious vomiting and a paucity of intestinal gas on abdominal x ray, had a midgut malrotation on contrast study. One child presented with a gastroschisis and one with a ruptured exomphalos, both had persistent abdominal distension and bilious vomiting during the postoperative period. After the 1st month of life, the most common mode of presentation was chronic constipation and abdominal distension with episodic vomiting. In three of the older children a febrile illness preceded the onset of gastrointestinal symptoms by a few days. Barium meal and follow through revealed a midgut malrotation in 16, a short small intestine in nine, and a non-hypertrophic pyloric stenosis in six. Six of these presented as neonates and had a midgut malrotation, short small intestine, and a non-hypertrophic pyloric stenosis. In four patients, three from one family, circular muscle morphogenesis was abnormal, with two circular muscle coats. In general, a barium enema was not helpful but suggested pseudo-Hirschsprung’s disease with a contracted left sided segment in 11 patients, nine of whom had nerve disease.

In those with an obstructed urinary system prenatally (n = 7), urinary tract infections were common. But in nine of 44, urinary tract anomalies were found on routine investigation without initial obstructive or infective symptoms. All of these had muscle disease.

FAMILY HISTORY

There were 11 cases in six families. In one family all three children were affected (female twins and their brother), in another two sisters, and in two families a brother and sister were
affected, suggesting an autosomal mode of inheritance. In the two remaining families (one with myopathic, the other with neuropathic disease) the inheritance appeared to be X-linked. We have shown previously that the latter condition maps to chromosome Xq28.14

ASSOCIATED ANOMALIES

There was a high incidence of associated anomalies, 16 of the patients had a midgut malrotation, 10 of whom had a shorter than normal small bowel. The urinary tract was involved in 16 of the patients.

HISTOPATHOLOGY

Histopathological evidence of enteric neuromuscular disease10121516 was seen in 38 of the 44 patients. Enteric muscle disease was seen in 22, including four patients with an additional smooth muscle coat. Intestinal neuropathy was present in 16.

MOTILITY STUDIES

Manometry was performed in 30 children. A myopathic trace was present in 13 and a neuropathic trace in 14 patients. In three children no contractile activity was seen. These three all had myopathic disease. The small intestinal manometry correlated well with the final histology.

Electrogastrography was carried out in 16 patients. In 10 of these patients, no dominant frequency could be found, which is suggestive of muscle disease; in five a tachygastria indicating a neuropathic process was found; and in one patient a normal dominant frequency of 0.048 Hz was present.

MEDICATION

Carbachol, bethanechol, propranolol, and metoclopramide were used up until 1984 with no consistent success. From 1984 cisapride was used in doses of 0.2–0.3 mg/kg body weight three times daily; this produced a temporary improvement in symptoms in six of 22 patients. Bacterial overgrowth was treated according to antibiotic sensitivity. Colistin, cotrimoxazole, and metronidazole were the most common antibiotics used for treating intestinal bacterial overgrowth. Antibiotics were helpful in reducing stool output and abdominal distension but only for short periods of time.

SURGICAL MANAGEMENT

The diagnosis of CIIPS was not considered at the initial laparotomy in 28 of 44 patients. In 10 patients, CIIPS was diagnosed on clinical history and on the results of investigations without resorting to surgery. Table 2 shows the outcome of the initial surgical procedures. There was no significant relief of symptoms following Ladd’s procedure, pyloromyotomy, pyloric excision, or gastro-jejunostomy. Colostomy effectively relieved symptoms in five of 11 patients, whereas in the remaining six patients the colostomy had to be converted to ileostomy because the obstructive symptoms persisted. A total of 31 patients had an ileostomy, with symptomatic relief in 16.

Stomal complications were common. Half of the children had an irregular ileostomy output alternating between low output with abdominal distension and vomiting followed by massive ileostomy losses resulting in fluid and electrolyte imbalance.

Eight children had a total of 17 ileostomy revision procedures, eight for prolapse of stoma and nine for suspected mechanical occlusion. Multiple adhesions were found at each operation but there were no correctable focal mechanical obstructing lesions and there was no noticeable change in ileostomy function after the revisional surgery. Four patients had their ileostomies closed when they were asymptomatic but all four needed replacement ileostomy as they rapidly developed acute obstructive symptoms.

Reconstitution of continuity of the gastrointestinal tract was carried out in seven patients after initial stoma formation, four with closure of colostomy and three by ileo-colic anastomosis. None of these procedures was successful. The three in whom an ileo-colic anastomosis was done required an ileo-rectal Duhamel pull through to relieve obstructive symptoms. Six children had an ileo-rectal Duhamel pull through with good outcome. The results of the various operative procedures are shown in table 3.

NUTRITIONAL SUPPORT

Thirty two patients required long term parenteral nutrition (TPN); 19 for less than six months, 11 of whom died. Thirteen needed TPN for longer than six months, 10 of these are still TPN dependent (for up to 16 years), and three are dead. Only eight patients have successfully been weaned off TPN, all of whom needed TPN for short periods only, with the maximum duration of less than six months.

OUTCOME

The outcome was defined as good when the child was alive and enterally fed and poor if the child had either died or was dependent on TPN. Of the 44 patients, 20 are alive and enterally fed and thus have a good outcome. However 14 have died and an additional 10 experience recurrent episodes of pseudo-

### Table 2 Primary surgical procedures and outcome

<table>
<thead>
<tr>
<th>Surgical procedure</th>
<th>Number of patients</th>
<th>Number with relief of symptoms</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ladd’s procedure</td>
<td>10</td>
<td>0</td>
</tr>
<tr>
<td>Pyloromyotomy/excision</td>
<td>5</td>
<td>0</td>
</tr>
<tr>
<td>Gastro-jejunostomy</td>
<td>2</td>
<td>0</td>
</tr>
<tr>
<td>Colostomy</td>
<td>11</td>
<td>5</td>
</tr>
<tr>
<td>Ileostomy</td>
<td>31</td>
<td>16 (10 total, 6 partial)</td>
</tr>
</tbody>
</table>

### Table 3 Definitive surgical procedures

<table>
<thead>
<tr>
<th>Procedure</th>
<th>Number of patients</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Colostomy closure</td>
<td>4</td>
<td>Constipation</td>
</tr>
<tr>
<td>Colorectal pull through</td>
<td>1</td>
<td>Constipation and soiling</td>
</tr>
<tr>
<td>Colorectal pull through</td>
<td>2</td>
<td>Needed ileo-rectal pull through</td>
</tr>
<tr>
<td>Ileorectal pull through</td>
<td>3</td>
<td>Needed ileo-rectal pull through</td>
</tr>
<tr>
<td>Ileorectal pull through</td>
<td>6</td>
<td>Very good</td>
</tr>
<tr>
<td>Ileoanal anastomosis</td>
<td>1</td>
<td>Very good</td>
</tr>
</tbody>
</table>
obstruction and are dependent on TPN. Therefore, these 24 patients had a poor outcome. Of the infants who died, three died as a result of withholding or withdrawing active treatment, nine died secondary to TPN related complications, and two died postoperatively after a laparotomy at other hospitals.

Fisher’s exact probability test was applied to a number of factors to determine which had a significant influence on the outcome. These factors and their significance are shown in table 4. The presence of midgut malrotation, short small intestine, involvement of the urinary system, <1 year of age at onset, and myopathy on histology were significantly (p < 0.05) poor prognostic factors. The sex of the child and a neuropathic histology were not significant features, with the exception of one child with acquired autoimmune aganglionosis who presented at the age of 10 years.

**Discussion**

CIIPS in children has been reported under a variety of names; megacystis-microcolon-intestinal hypoperistalsis syndrome, intestinal pseudo-obstruction, chronic adynamic ileus, pseudo-Hirschsprung’s disease, adynamic bowel syndrome, colonic neuronal dysplasia, and hollow visceral myopathy. It is the result of a heterogeneous group of disorders of the enteric neuromusculature that cause severe distension associated with obstruction and episodic vomiting, which may be bile stained. Previous studies have either been of small numbers of patients or have been multicentred with poorly validated data.

The diagnosis of CIIPS is dependent on the awareness of the clinician, the recognition of the clinical syndrome, and the exclusion of mechanical obstruction by radiological assessment and exclusion of Hirschsprung’s disease by rectal biopsy. The patient often undergoes exploratory laparotomy before the diagnosis is considered. The main diagnostic difficulties are the poor specificity of clinical features and the absence of established diagnostic tests.

**Table 4 Prognostic factors**

<table>
<thead>
<tr>
<th></th>
<th>Total</th>
<th>Good outcome</th>
<th>Poor outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Malrotation</td>
<td>16</td>
<td>3</td>
<td>13</td>
</tr>
<tr>
<td>Short small bowel</td>
<td>9</td>
<td>1</td>
<td>8</td>
</tr>
<tr>
<td>Urinary involvement</td>
<td>16</td>
<td>4</td>
<td>12</td>
</tr>
<tr>
<td>Muscle disease</td>
<td>22</td>
<td>7</td>
<td>15</td>
</tr>
</tbody>
</table>

p < 0.05 for all prognostic factors.

The presence of midgut malrotation, short small intestine, involvement of the urinary system, <1 year of age at onset, and myopathy on histology were significantly (p < 0.05) poor prognostic factors. The sex of the child and a neuropathic histology were not significant features, with the exception of one child with acquired autoimmune aganglionosis who presented at the age of 10 years.

The initial objectives of treatment must be to rehydrate and restore the normal electrolyte and acid/base balance, to control obstructive symptoms, and to control urinary and enteric bacterial overgrowth. This will require medical, surgical, and nutritional management. Each has a part to play and must be used in the correct manner for optimal treatment.

Carbachol, bethanocol, propranolol, metoclopramide, erythromycin, and neostigmine have all been used to restore the disturbed gastrointestinal motility in patients with CIIPS, but with only limited success. Cisapride has been shown to be the most effective prokinetic agent. In our series, cisapride achieved only temporary relief of symptoms in about a quarter of patients and the symptoms returned within a few months in all.

Treatment of bacterial overgrowth with broad spectrum antibiotics was difficult, and was only successful when microbiology isolated the important dominating organisms of the intestinal flora. Urinary infections were con-
Chronic intestinal pseudo-obstruction

Key messages
- Chronic intestinal pseudo-obstruction requires coordinated investigation by physician, surgeon, and pathologist
- Manometry and histopathological diagnosis provide prognostic information
- Decompression stomas provide treatment, allow diagnosis, and may prevent unnecessary further surgery
- Long term total parenteral nutrition may be required

trolled using standard treatments for recurrent infections.

Until now, the place of surgery in the management of CIIPS has been ill defined. However, our study clearly shows that pyloromyotomy, excision of the pylorus, and gastrojejunostomy are of no benefit in relieving symptoms because the apparent gastric outlet obstruction seen in this disorder is secondary to dysmotility of the stomach and intestine. Ladd’s procedure for malrotation was similarly unsuccessful in relieving the obstructive symptoms of the underlying enteric neuromuscular disease, but did prevent volvulus from occurring.

Dramatic symptom relief can be expected with a defunctioning stoma if the pathology is confined to the bowel distal to the stoma. In our series a defunctioning stoma relieved symptoms completely in a quarter of the patients in whom the disorder was confined to the terminal ileum and colon. It was partially successful in a quarter and was ineffectual in half of patients in whom the gut was diffusely affected along its entire length. Vargas et al reported the results of a large survey in North America conducted in 1988 in which patients were reported by questionnaire. Exact numbers of patients who underwent surgery were not given but they felt that in many of the patients diversion through defunctioning stomas was done with temporary, partial, or no relief of obstruction. Another recent review of children who had defunctioning stomas showed no relief of symptoms. However, like our present study other reports do show benefit from defunctioning or decompression stomas. Similarly, the effectiveness of a stoma in reducing gaseous distention and bloating in patients on parenteral nutrition has been reported previously. It is advantageous to decompress the small bowel early in the course of the illness, before gross dilatation occurs, because this itself reduces effective motor activity of the gut. In our series, meticulously performed end ileostomy reduced the chances of stoma prolapse and further simplified management.

Even with a functioning ileostomy, episodes of functional obstruction occurred in our patients. Typically, the ileostomy output was irregular, with periods of massive output alternating with periods of no output. Paradoxically, bilious nasogastric aspirates coexisted with large ileostomy losses. There should be unequivocal evidence of mechanical obstruction before performing any further laparotomies, not least because our children were prone to prolonged paralytic ileus after any surgery, and at laparotomy multiple adhesions were always found. Further surgery can only add to them. Our experience in this series amply supports earlier studies that pointed out that repeated surgery only resulted in dense adhesions and increased morbidity.

High ileostomy washouts were effective in two of our patients. The success of this treatment in these patients might have been the result of less extensive disease or the efficacy of daily ileostomy washouts in allowing regular decompression of the small bowel. It may be worthwhile to evaluate this procedure further to determine its efficacy.

Once a defunctioning stoma has relieved the symptoms the next inevitable question is whether the stoma can be dispensed with. In our series, four children with well functioning ileostomies had their stomas closed; however, this was unsuccessful and all developed acute obstructive symptoms and needed ileostomies to be refashioned. The cause of failure in these cases was persistent dysmotility of the bowel proximal to the stoma. An ileo-rectal Duhamel pull through procedure using the ileum proximal to the stoma proved to be the best definitive procedure in our series. It would appear logical to bypass all the affected bowel similar to the definitive management of Hirschsprung’s disease; however, this might not always be possible without resecting excessive amounts of intestine. Although it may appear illogical to place a stoma in a bowel similar to the definitive management of Hirschsprung’s disease; however, this might not always be possible without resecting excessive amounts of intestine. Although it may appear illogical to place a stoma in a bowel similar to the definitive management of Hirschsprung’s disease; however, this might not always be possible without resecting excessive amounts of intestine.

In children with extensive severe disease TPN has been the mainstay of treatment. If there are no signs of improvement in intestinal function within a few weeks, a tunnelled central venous catheter should be inserted surgically, because it is likely that the need for TPN will continue for many months. Catheter blockage and/or infections are the main complications associated with long term TPN. Before 1987, seven children died while on TPN for less than one year because of TPN related complications. However, with meticulous nursing care or with “home TPN” the incidence of line infections has since been lower. The duration of dependence on TPN appears to be of crucial prognostic importance. There have been 10 survivors among the 13 patients requiring TPN for longer than six months. All 10 have failed repeated attempts to wean them off parenteral nutrition. The only seven children currently on full enteral nutrition required TPN for less than six months. It appears that if the disease is severe enough to require TPN for more than six months, it is likely that the child will be dependent on TPN for at least four years.

The above data suggest that with extensive severe disease only a small improvement in bowel motility with the passage of time can be...
anticipated, as has been proposed previously in some adults.14 Patients with an acquired disease, particularly of autoimmune origin, may deteriorate without specific treatment.14 One such patient with an autoimmune ganglionitis succeeded in denervating her entire gastrointestinal tract. Ultimately, there were serious vascular access problems and she underwent intestinal transplantation. She is well now some five years after transplantation. Recent advances in enteral nutrition, such as modular feeds and continuous rate infusions, have been helpful in weaning children off TPN.16 In nine children in our series, TPN was completely avoided by the combination of dietary measures and a defunctioning stoma.

We were able to identify five poor prognostic factors: involvement of the urinary tract, early age at onset of symptoms, short small intestine, midgut malrotation, and myopathic histology. The latter three are known to be common in severe disease,56 57 and thus might be expected to result in poor prognosis. The only surgical procedures that were successful were Ladd’s procedure where there was a malrotation and this prevented intestinal volvulus, a defunctioning ileostomy, and ultimately an ileo-rectal Duhamel pull through, especially where the disease was confined to the terminal ileum.

Duhamel pull through, especially where the disease was confined to the terminal ileum and colon. Our observations at surgery confirmed the propensity for patients with these disorders to form dense adhesions postoperatively. From our studies, we would strongly suggest that only planned surgery for initial diagnosis and bowel decompression should be undertaken in these patients. Laparotomies for obstruction should only be performed where there are clear focal signs or mechanical obstruction has been demonstrated.

References


28 Ament ME. Chronic intestinal pseudo-obstruction syndrome in pediatrics: results of a national survey by members of the North American Society of Pae...


Chronic intestinal pseudo-obstruction: treatment and long term follow up of 44 patients

S Heneyke, V V Smith, L Spitz and P J Milla

Arch Dis Child 1999 81: 21-27
doi: 10.1136/adc.81.1.21

Updated information and services can be found at:
http://adc.bmj.com/content/81/1/21

These include:

References
This article cites 46 articles, 7 of which you can access for free at:
http://adc.bmj.com/content/81/1/21#BIBL

Email alerting service
Receive free email alerts when new articles cite this article. Sign up in the box at the top right corner of the online article.

Topic Collections
Articles on similar topics can be found in the following collections

Childhood nutrition (712)
Childhood nutrition (paediatrics) (396)
Muscle disease (62)
Neuromuscular disease (166)
Rheumatology (521)
Small intestine (46)
Drugs: CNS (not psychiatric) (497)
Pathology (248)
Gastric emptying disorders (12)
Oncology (777)
Stomach and duodenum (55)

Notes

To request permissions go to:
http://group.bmj.com/group/rights-licensing/permissions

To order reprints go to:
http://journals.bmj.com/cgi/reprintform

To subscribe to BMJ go to:
http://group.bmj.com/subscribe/