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

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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).

(Arch Dis Child 1999;81:21–27)

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 the last 10 years few patients survived long enough for the natural history of CIIPS to be apparent, and the place of surgery in its management is still not well defined. The aims of our study were to document the course of the disorder in a large number of patients, in whom the underlying enteric neuromuscular disease had been clarified, from one centre over a long period of time, to determine the place and type of surgery used in their management, and to identify prognostic factors.

The histopathological features and motility abnormalities have been reported previously and will not be considered in detail here.

Methods

Between 1979 and 1997, 44 children with recurrent symptoms of bowel obstruction in the absence of a mechanical occlusion, in whom Hirschsprung’s disease was excluded, were investigated prospectively to define the cause and extent of the bowel disorder.

We noted the age at onset of illness, the presenting symptoms, drugs given, specific nutritional support used, and the types of surgery performed. We performed marker transit studies, abdominal ultrasound, and contrast x ray studies in all children; upper gastrointestinal contrast studies in 32 children, and contrast studies of the lower gastrointestinal tract in 22. If there was involvement of the urinary tract, the extent of involvement, the management, and the outcome were noted.

HISTOPATHOLOGY

Full thickness intestinal biopsies were available from 44 patients, but for two patients the material was obtained postmortem. These comprised both small and large intestine from 22 patients, small bowel alone from 17, and large bowel alone from five. We performed routine histology, enzyme histochemistry, and immunocytochemistry by light microscopy and ultrastructural studies by electron microscopy as described previously (VV Smith, PhD thesis, University of London, 1993).10–12

MOTILITY STUDIES

Surface electrogastrography

We recorded fasting gastric electrical control activity (ECA) in 16 patients who initially presented after 1989 and the results were analysed using our previously described method. In summary, four pairs of Ag–AgCl electrodes matched for impedance were placed along the greater curvature of the stomach from the fundus to the first part of the duodenum and the potential differences across these pairs of electrodes were measured for one hour after a four hour fast or during small intestinal manometry. The frequencies present were determined by autoregressive modelling using a running spectral analysis method. After running spectral analysis of gastric ECA the dominant frequency was determined if present.
**Table 1 Clinical details of 44 patients with chronic intestinal pseudo-obstruction**

<table>
<thead>
<tr>
<th>Patients</th>
<th>Birth</th>
<th>Present</th>
<th>Weight centile</th>
<th>Symptoms</th>
<th>Radiology</th>
<th>Histology</th>
<th>Surgery</th>
<th>Outcome</th>
</tr>
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<tbody>
<tr>
<td>1–6</td>
<td>3rd to 50th</td>
<td>Birth</td>
<td>–</td>
<td>Vomiting</td>
<td>Dilated whole gut</td>
<td>5 M</td>
<td>6 Ladd’s procedure</td>
<td>4 dead</td>
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<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Intestinal obstruction</td>
<td>Pyloric obstruction</td>
<td>1 N</td>
<td>6 ileostomy</td>
<td>2 TPN</td>
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<td></td>
<td></td>
<td></td>
<td></td>
<td>Short small intestine</td>
<td>Malrotation</td>
<td>6 pyloromyotomy</td>
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<tr>
<td>7–13</td>
<td>3rd to 25th</td>
<td>Antenatal</td>
<td>–</td>
<td>Failure to pass urine</td>
<td>Dilated bowel</td>
<td>M</td>
<td>Ileostomy</td>
<td>4 dead</td>
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<td></td>
<td></td>
<td></td>
<td></td>
<td>Intestinal obstruction</td>
<td>Megacystis</td>
<td>1 enteral</td>
<td></td>
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<td></td>
<td></td>
<td></td>
<td></td>
<td>Megacystitis</td>
<td>Megaoatere</td>
<td>2 TPN</td>
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<td></td>
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<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>Dilated bowel</td>
<td>9 M</td>
<td>Ileostomy</td>
<td>3 dead</td>
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<td></td>
<td>Megacystis</td>
<td>Megaoatere</td>
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<td></td>
<td>Abdominal distension</td>
<td>Slow transit</td>
<td>2 TPN</td>
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<td></td>
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<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>Dilated bowel</td>
<td>6 N</td>
<td>colostomy</td>
<td>2 gastrostomy</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>1 stricture</td>
<td>Slow transit</td>
<td>4 normal feeds</td>
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<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>Dilated bowel</td>
<td>10 contracted colon</td>
<td>8 N</td>
<td>colostomy</td>
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<td></td>
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<td></td>
<td></td>
<td>Obstruction</td>
<td>3 malrotation</td>
<td>2 ileostomy</td>
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<td>Abdominal distension</td>
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<td>43</td>
<td>10th</td>
<td>&lt; 3rd</td>
<td>2 years</td>
<td>Obstruction</td>
<td>Dilated bowel</td>
<td>M</td>
<td>Ileostomy</td>
<td>TPN</td>
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<td>44</td>
<td>25th</td>
<td>&lt; 3rd</td>
<td>10 years</td>
<td>Severe constipation</td>
<td>Dilated bowel</td>
<td>N</td>
<td>Ileostomy</td>
<td>TPN</td>
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<td>Abdominal distension</td>
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**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.13 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.13 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 disease10 12 15 16 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-jejunalostomy. 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-
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.33

**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.14–23 It is the result of a heterogeneous group of disorders of the enteric neuromusculature that cause severe intestinal dysmotility, resulting in functional obstruction.24–30

Analysis of published studies has shown that most patients develop symptoms early in life.31 In this series, 32 of 44 children developed symptoms during the 1st year of life. At birth they may present with obstruction of either the gut or urinary tract, or both. A persistent bilious aspirate can indicate an associated midgut malrotation. Later, the main presentation is with chronic constipation and/or abdominal 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.14–32

<table>
<thead>
<tr>
<th>Prognostic factors</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.*

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CIIPS should be considered as a probable diagnosis when there are continuing symptoms after Ladd’s procedure for midgut malrotation, when intestinal obstruction is associated with bladder dysmotility, or when in early life there is recurrent obstruction after exclusion of Hirschsprung’s disease by rectal biopsy. In our hands, surface electrogastrography has proved to be a useful screening investigation followed by upper gastrointestinal contrast study. Dilated small bowel with impaired or retrograde peristalsis and stasis of contrast are suggestive of small bowel dysmotility,33 which requires confirmation by antrudodenal manometry. In addition, it may differentiate between a myopathic process and a neuropathic process.34–35

The aim should be to avoid an exploratory laparotomy but to have planned surgery if there is a need for a defunctioning ileostomy to decompress the intestine. At this time, it is mandatory that full thickness tissue is obtained to determine the type of enteric neuromuscular disease. Our data indicate that this has prognostic value, with muscle disease having a poorer outcome than nerve disease, and the presence of muscle disease may indicate a congenital disorder. Because of the subtle nature of abnormalities in enteric nerves and muscle in CIIPS, routine microscopy alone is generally unhelpful in making the diagnosis. The use of electron microscopy and histochemical (enzyme and immunohistochemistry) techniques allows the detection of a wider range of changes, thus enabling a more accurate histological classification of the patients into myopathic and neuropathic groups,32 which is relevant in predicting the clinical outcome. Therefore, it is important to preserve the resected full thickness sample in ways that permit ultrastructural examination (glutaraldehyde fixation), immunohistochemistry, and enzyme histochemistry (snap freezing), in addition to routine light microscopy (VV Smith, PhD thesis). (Although silver staining was used in the early patients (17 of 44) in our series, it must be born in mind that it is unreliable in young babies because argyrophilic neurons are present constantly only in children older than 1 year,36 and we feel that this method is no longer of value in the paediatric population.)

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.37–40 Cisapride has been shown to be the most effective prokinetic agent.31–34 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

25

relief of obstruction.44

mas was done with temporary, partial, or no

patients diversion through defunctioning sto-

been reported previously.48 49 It is advantageous

bloating in patients on parenteral nutrition has

stoma in reducing gaseous distension and

occurs, because this itself reduces e

course of the illness, before gross dilatation

to decompress the small bowel early in the

symptoms of the underlying enteric neuromuscular
disease, but did prevent volvulus from occur-

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.44 Exact num-

bers of patients who underwent surgery were

not given but they felt that in many of the

patients diversion through defunctioning stoma-

was done with temporary, partial, or no

relief of obstruction.44 Another recent review of

children who had defunctioning stomas

showed no relief of symptoms.45 However, like

our present study other reports do show benefit

from defunctioning or decompression

stomas.46 47 Similarly, the effectiveness of a

stoma in reducing gaseous distension and

bloating in patients on parenteral nutrition has

been reported previously.46 47 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, meticu-
lously performed end ileostomy reduced the

chances of stoma prolapse and further simpli-

fied 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 alter-

nating with periods of no output. Paradoxi-

cally, bilious nasogastric aspirates coexisted

with large ileostomy losses. There should be

unequivocal evidence of mechanical obstruc-
tion before performing any further laparoto-

mies, 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 sup-

ports earlier studies that pointed out that

repeated surgery only resulted in dense adhe-

sions and increased morbidity.44

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 proxim-

tal to the stoma proved to be the best defini-
tive procedure in our series. It would appear

logical to bypass all the affected bowel similar
to the definitive management of Hirsch-

sprung’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 affected bowel,

there may be sufficient residual motor function

to allow the stoma to act but not to evacuate

the bowel through the anus.

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.52 53 After eight years

of experience, 52 53 54 55 Catheter blockage and/or
infected lines 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 meticu-
lous nursing care or with “home TPN” the

incidence of line infections has since been

lower.52 53 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 nutri-
tion 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

Dr. John Doe

March 20, 2023

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
anticipated, as has been proposed previously in
some adults.45 Patients with an acquired
disease, particularly of autoimmune origin,
might deteriorate without specific treatment.46
One such patient with an autoimmune ganglio-
nosis 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.47 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 myopathy 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 defunction-
ing ileostomy, and ultimately an ileo-rectal
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.

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**FETAL AND NEONATAL EDITION**

**July issue**

The following articles—being published in the July 1999 issue of the *Fetal and Neonatal* edition of the *Archives of Disease in Childhood*—may be of general interest to paediatricians.

**ANNOTATION**

Recent advances in neonatology
J M Rennie, S A Bokhari

**ORIGINAL ARTICLES**

Neurodevelopmental outcome at early school age of children born to mothers with gestational diabetes
A Ornoy, A Wolf, N Ratzen C Greenbaum, M Dulitzky

Iron nutritional status in preterm infants fed formula fortified with iron
Ian J Griffin, Richard J Cooke, Michael M Reid, Kenneth PB McCormick, Jacqui S Smith

Longitudinal study of behaviour disorders in low birthweight infants
C J Stevenson, P Blackburn, P O D Pharoah

Safety and effectiveness of BCG vaccination in preterm babies
Sudhin Thayyil-Sudhan, Ashok Kumar, Mehbar Singh, Vinod Kumar Paul, Ashok Kumar Deorari

Severe apnoeas following immunisation in premature infants
M H Slack, D Schapira

**CURRENT TOPIC**

Low birth weight and adult insulin resistance: the “catch-up growth” hypothesis
Stefano Gianfarani, Daniela Germani, Francesco Branca
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
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