Chronic Idiopathic Intestinal Pseudo-obstruction

A Surgical Approach

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Chronic idiopathic intestinal pseudo-obstruction is an increasingly recognized syndrome in which patients usually present with an acute or chronic history suggestive of intestinal obstruction, although no obstructing lesion is found at surgery. The diagnosis can be suspected in most cases from the clinical presentation. A diagnostic evaluation should be undertaken and exploratory laparotomy avoided if the diagnosis is confirmed on the basis of the radiographic and manometric data. If, in the acute presentation, exploratory laparotomy proves unavoidable, and dilated, nonmechanically obstructed bowel is found, a full-thickness biopsy specimen should usually be taken and the abdomen closed. A carefully chosen, palliative procedure should be reserved for patients who have well defined clinicoanatomic patterns of involvement, and who are incapacitated by their symptoms despite medical management.

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INTESTINAL PSEUDO-OBSTRUCTION is a clinical syndrome characterized by signs and symptoms of mechanical obstruction without a mechanical origin present.[4,14] The most common type of pseudo-obstruction is the acute variety, which has many synonyms—paralytic ileus, postoperative ileus and spastic ileus. Acute pseudo-obstruction is usually self-limited and related to operative procedures or acute illnesses, such as pancreatitis, cholecystitis, pneumonia or myocardial infarction. Chronic intestinal pseudo-obstruction is much less common and, although it may be secondary to a known systemic disease, such as progressive systemic sclerosis or amyloidosis, it is often idiopathic. In the latter case it is called chronic idiopathic intestinal pseudo-obstruction (CIIP).

Regardless of cause of CIIP, its pathophysiology involves loss of coordinated intestinal propulsive motility, often throughout the gastrointestinal tract.[4] Although no identifiable disease is present in some patients,[9,21,27] the smooth muscle or myenteric plexus contain morphologic abnormalities in most patients. In addition, many cases are familial. Hereditary hollow visceral myopathy and familial visceral myopathy are two terms which delineate a form of CIIP caused by smooth muscle degeneration and which is transmitted as an autosomal dominant trait.[5,23] Two other forms of CIIP are caused by degeneration of the myenteric plexus. One form of CIIP is familial, involves the central and peripheral nervous systems as well as the intestinal nervous system, and is marked by the presence of an eosinophilic intranuclear inclusion.[22] The other form is not familial, involves only the intestinal nervous system, and intranuclear inclusions are absent.[3] The former is called familial visceral neuropathy and the latter is called sporadic visceral neuropathy.

Medical management of CIIP is difficult because there is no drug which can restore normal propulsive motility. Anorexia, vomiting, and malabsorption are common and lead to such significant weight loss that some patients must be placed on home parenteral nutrition.

Many patients undergo laparotomy either because CIIP is not diagnosed or because a palliative procedure is attempted.[4,12,14] Despite the fact that these patients often undergo surgery, little is written about CIIP in the surgical literature. The purpose of this

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paper is to review our experience with 15 patients having CIIP. From this experience and a review of the literature, we suggest some guidelines which, one would hope, will help in the surgical management of this difficult syndrome.

Patient Population and Case Histories

Thirty patients with chronic intestinal pseudo-obstruction were studied between January 1972 and November 1979. Of these, 14 patients had scleroderma and one patient had sclerosing mesenteritis. The other 15 patients had various types of CIIP and are the subjects of this report. The diagnosis of CIIP was made in patients with no recognized underlying disease, who presented with chronic obstructive symptoms. The possibility of mechanical obstruction was ruled out by surgical, autopsy or radiographic examinations. Radiographs of the small bowel demonstrated dilatation of the duodenum and/or small intestine in all patients, and of 13 patients who had esophageal manometric evaluation, 11 patients had aperistalsis.

In eight of the 15 patients, adequate tissue was available for detailed histologic study. Five patients had hollow visceral myopathy[23]; two patients had familial visceral neuropathy,[22] and one patient had sporadic visceral neuropathy.[3] Of the other seven patients, one had diffuse jejunal diverticulosis and the other six patients had either no tissue or insufficient tissue available to make a specific pathologic diagnosis.

Despite the various histologic factors, the clinical picture was similar for the 15 patients. The duration of obstructive symptoms varied from two to 57 years. All patients experienced recurrent episodes of vomiting and 14 of these 15 patients had attacks of abdominal distension and pain. Abdominal distention was sometimes equal to the size of a full term pregnancy. Ten patients had diarrhea and 11 patients had constipation; any patient could have alternating diarrhea and constipation, or diarrhea during one period of life and constipation in another. Six patients had dysphagia, whereas only one had pyrosis. Two patients had symptoms of urinary retention from concomitant bladder involvement.

Physical findings included abdominal distention, in nine of 13 patients, and succussion, in seven of nine patients. The patients weighed an average of 43.7 kg (range: 22–59 kg except for 1 patient who weighed 83 kg). Excluding this latter patient, all the patients lost an average weight of 21 kg (range: 6–48 kg).

Although most patients presented with multiple gastrointestinal symptoms, it was often possible to identify one or two dominant complaints in individual patients. By correlating the dominant complaints with

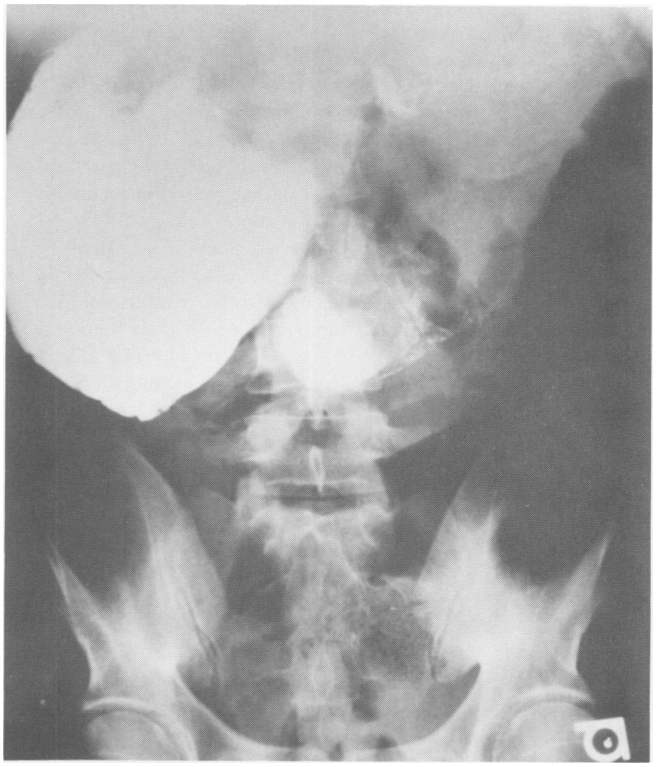

FIG. 1. Dilated, atonic duodenum, characteristic of gastroduodenal predominence (case 4).

radiographic and surgical findings, we identified four clinicoanatomic patterns of CIIP. These patterns were 1) esophageal, in which dysphagia predominated; 2) gastroduodenal, in which a clinical syndrome of duodenal or gastroduodenal obstruction predominanted as characterized by nausea, vomiting, abdominal pain, and usually mild abdominal distension; the upper gastrointestinal series usually demonstrated a megaduodenum (Fig. 1), sometimes in association with dilated stomach; 3) small bowel, in which moderate to marked abdominal distension was associated with pain and vomiting. Plain abdominal radiographs showed dilated loops of small intestine and radiographs of the small bowel demonstrated diffuse intestinal dilatation with or without a megaduodenum; 4) colonic, in which abdominal distension and discomfort were associated with constipation that sometimes alternated with diarrhea. The passage of large amounts of flatus and stool partially relieved these complaints and was associated with a diminution of abdominal distention. Radiographs demonstrated grossly distended loops of colon with variable amounts of distended small bowel.

Although many of these symptoms were present to some degree in every patient, only one or two symptoms tended to predominate in individual patients. The following case histories illustrate examples of these patterns.

Esophageal Predominance

This 26-year-old man was well until the age of 24 years, when he developed recurrent abdominal pain, bloating to the size of a three-month pregnancy, and vomiting. The patient's food intake decreased by more than 50% and his weight decreased by 14 kg over a 2-month period. Although he initially had one stool per day, he developed increasingly severe constipation and required laxatives. Barium contrast radiographs of the upper and lower intestines demonstrated a questionable duodenal ulcer. After being hospitalized four times in two months, the patient was referred to the authors for evaluation. Neither the patient's parents nor his two siblings has any gastrointestinal symptoms. Repeat radiographs revealed a normal esophagus and stomach. However, a megaduodenum was present and the colon was elongated, redundant, and emptied poorly. The small intestine distal to the duodenum was normal. Esophageal manometric studies revealed normal peristalsis, a lower esophageal sphincter (LES) pressure of 12 mmHg, normal LES relaxation, and acid reflux on loading the stomach with 300 ml of HCl. A cystometrogram was normal.

Although no specific treatment was given, the patient's symptoms gradually improved and his weight increased to 60 kg. Several months later, however, he developed dysphagia for liquids and solids, which occasionally was associated with chest discomfort or pain. He had daytime and nocturnal regurgitation of undigested food and awakened with nocturnal coughing. He also developed foul smelling diarrhea which resolved after the administration of antibiotics. Although abdominal pain and distention still occurred occasionally, he identified dysphagia and regurgitation as his most troublesome symptoms. Radiographs (taken 13 months after the initial studies) showed a dilated esophagus which had a fluid level at 17.5 cm above the gastroesophageal junction. Peristalsis was absent in the lower two-thirds of the esophagus and there was a large amount of retained food. The stomach was mildly dilated, contained old food, and emptied slowly. Esophageal manometric studies showed normal peristalsis in the upper esophagus. Peristalsis in the lower two-thirds was absent and swallows elicited low amplitude, simultaneous waves. Swallows were sometimes associated with a rise of baseline pressure and four to five consecutive waves. The LES pressure was 35 mmHg and only intermittently relaxed to intragastric pressure. There was no esophageal reflux.

The patient underwent balloon dilatation of the gastroesophageal junction which produced only a short-lived improvement. The dilatation was repeated two weeks later, following which, his symptoms markedly improved. The patient now has only mild dysphagia; there is no food retention and no regurgitation or nocturnal coughing. Unfortunately, the follow-up period has only been three months.

Comment. This patient went from no esophageal involvement to disabling involvement from functional esophageal obstruction. He has thus far done better after balloon dilatation.

Duodenal Predominance

This 20-year-old woman's (case 3) symptoms began in childhood, consisting initially of abdominal pain and bloating. Diarrhea developed later, but the patient was minimally symptomatic until age 15 years when she developed progressively severe abdominal distention and pain, and was finally hospitalized after two days of vomiting and obstipation. Radiographs showed markedly dilated loops of intestine, and an exploratory laparotomy revealed the entire small bowel and colon to be filled with air and fluid. No point of mechanical obstruction was identified and the incision was closed. Following discharge from the hospital, the patient's symptoms recurred and she was referred to the authors for evaluation.

Physical examination revealed a very thin, pale adolescent girl. Her abdomen was distended and hypertympanitic, and a succussion was audible. Bowel sounds were present with occasional rushes. Radiographic examination demonstrated esophageal aperistalsis, a normal stomach, a massively dilated duodenum, a less dilated small intestine distal to the duodenum, and an elongated, redundant, ahaustral colon. Esophageal manometric studies revealed absent peristalsis; swallows elicited low amplitude, simultaneous contractions. The LES pressure was 41 mmHg and it only partially relaxed following swallows.

The patient was treated with total parenteral nutrition and nasogastric suction. When her abdominal pain and vomiting had not resolved after two months, a retrocolic isoperistaltic gastrojejunostomy and Stamm gastrostomy were performed. The patient was still unable to eat two months later, and was miserable from persistent abdominal pain, distention and vomiting. Careful questioning convinced us that most of her discomfort was probably due to the distended duodenum. At re-exploration, the deuodenum was patulous and atonic and measured 10 cm in diameter. The gastrojejunostomy was widely patent and the distal jejunal loops were dilated. The gastrojejunostomy was taken down and converted to a Roux-en-Y. In addition, a 14 cm elliptical segment of the anterior duodenal wall was excised. The patient gradually improved and was discharged from the hospital six weeks later.

The patient has had follow-up examinations for 5 years. Although she has had intermittent attacks of abdominal pain and distention and has required occasional antibiotics to treat malabsorption, she has not been admitted to the hospital. She is maintaining her weight at 45 kg and is working full-time.

Comment. This patient's duodenum was acting as a large, poorly draining reservoir. The patient's symptoms were ameliorated by reducing its size and draining it more dependently into the jejunum.

Small Bowel Predominance

This 67-year-old man (case 8) first developed episodes of postprandial abdominal fullness and nausea at age 37 years, which was relieved by vomiting and belching and which were often followed by diarrhea. These episodes continued for the next 12 years at which time he developed severe abdominal pain which lead to an exploratory laparotomy. No specific disease was identified although the small intestine was thought to be rotated on itself. One year later, following a nonvisualising oral cholecystogram, a normal gall bladder was excised. A recurrent "obstructive" attack led to a diagnosis of small bowel obstruction two years later. At laparotomy, no point of obstruction was identified, but the jejunum was found to be greatly dilated and to have multiple diverticula, whereas the ileum was normal. Ninety centimeters of jejunum were resected and the small intestine was plicated.

Over the next several years the patient lost 12 kg in weight, and developed persistent steatorrhea and peripheral edema. On the authors' initial evaluation the patient was a pale, thin man who appeared relatively well. His abdomen was moderately distended and a succussion was audible. Pretibial edema was present and the neurologic examination was normal. Fat absorption was 57% of intake (normal: 93%) before the administration of antibiotics and 80% after. Vitamin B_{12} absorption was abnormally low. It did not improve with the administration of intrinsic factor, but did improve with the administration of antibiotics. Fluid aspirated from the jejunum prior to the administration of antibiotics cultured

a large number of *E. coli* and anaerobes. Radiographic studies showed a normal esophagus, stomach, and colon. The duodenum, jejunum, and proximal ileum were dilated but no diverticula were identified. Esophageal manometric studies demonstrated normal peristalsis.

The patient did well on intermittent administration of antibiotics and vitamin supplements for several years. He was eventually lost to follow-up study and presented elsewhere four years later with an acute exacerbation of obstructive symptoms. He did not inform the surgeon that he had pseudo-obstruction and an exploratory laparotomy was undertaken. As before, no mechanical obstruction was found and the patient was closed, only to return to the operating room several hours later to have a bleeding vessel ligated.

The patient was referred back to the University of Washington. The administration of antibiotics and liquid formula supplements did not improve the patient's condition and he had to be placed on home parenteral nutrition. The patient has regained his lost weight and has felt much stronger. However, he continues to intermittently vomit when he eats.

Comment. This case illustrates the lack of response to small intestinal resection and the needless operations resulting from misdiagnosis.

Colonic Predominance

This 64-year-old white man (case 7) had a 50-year history of intermittent episodes of abdominal distension and pain relieved by the passage of flatus. Until age 61 years he had one formed stool a day, but since then has had progressive constipation requiring laxatives. At age 62 years the patient first began to experience

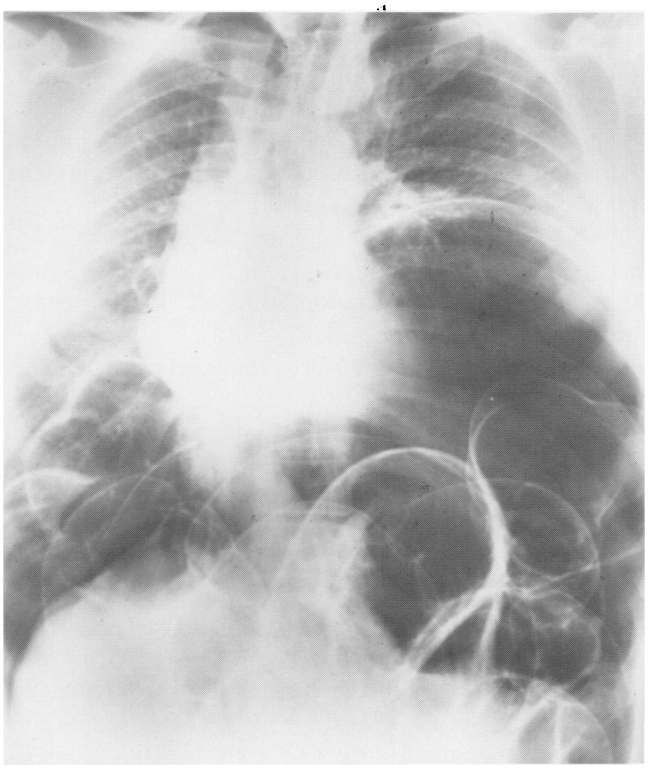

FIG. 2. Markedly dilated loops of small and large bowel. Free peritoneal air is also present although the patient had no peritoneal signs.

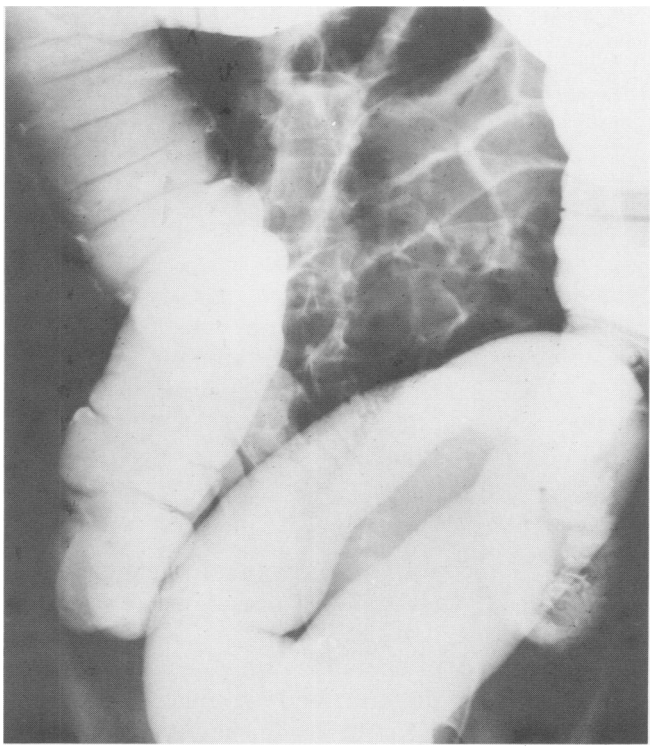

FIG. 3. The colon is markedly elongated and redundant, such that only a part of it could be seen on any one film.

epigastric discomfort, partially relieved by self-induced vomiting. For the three months prior to admission to the hospital the episodes of abdominal distension became more severe and frequent. During this period the diagnosis of CIIP was made on the basis of abnormal esophageal peristalsis and abnormal radiographic studies of the gastrointestinal tract. At admission to the hospital his abdominal girth was 43 inches, compared with a usual girth of 26 inches. The patient's main complaint was of increased shortness of breath and abdominal distension diminished by the passage of flatus. Physical examination demonstrated an emaciated man who weighed 65 kg. Physical findings included, jugular venous distention, bibasilar pulmonary rales, and massive lower extremity edema. The patient's abdomen was massively distended and hypertympanitic, and a succussion was present. Bowel sounds were present and peritoneal signs were absent. Laboratory data included a serum albumin level of 3.2 g/dl Radiographs of the chest demonstrated markedly dilated loops of large and small intestine, and a pneumoperitoneum which produced severe elevation of both diaphragms with displacement of mediastinal structures to the right (Fig. 2). Four liters of barium contrast material were required to fill a grossly dilated, elongated, and redundant colon (Fig. 3), and barium studies showed a mildly dilated small intestine. Pulmonary function tests revealed a moderate restrictive defect with a decreased diffusing capacity of 42%, suggesting maldistribution of ventilation. Esophageal manometric studies demonstrated aperistalsis, low amplitude simultaneous waves after swallows, an LES pressure of 10 mmHg, normal LES relaxation, and spontaneous acid reflux.

Medical management of the patient's abdominal and respiratory symptoms was unsuccessful. At operation a grossly hypertrophied and elongated stomach, duodenum, and proximal jejunum were found. The remainder of the jejunum and main portion of ileum were normal. The terminal ileum and the entire abdominal colon

were grossly dilated, redundant, and thickened. The terminal ileum and colon were resected, and an end-to-side ileosigmoidostomy was performed at the level of the peritoneal reflection.

Postoperative, the patient's respiratory condition improved, and over a one-week period, he diuresed 10 kg with resolution of the lower extremity edema. Since being discharged from the hospital the patient has continued to experience some abdominal discomfort but remains significantly improved.

Comment. This patient suffered severe cardiopulmonary embarrassment from massively dilated intestine associated with severe constipation. The colectomy resulted in two to three stools a day and the ability to pass flatus easily. This relieved the pressure on the patient's diaphragms and ameliorated his cardiopulmonary symptoms.

Results

Seventy-three procedures were performed on 12 patients, with a range of 1–34 operations per patient (Table 1).

A wide variety of operations were performed, making it difficult to discern trends in clinical response. Several comments can be made, however, from the surgical data.

1) A review of the operative notes from the initial procedures revealed that in 11 of 12 cases, CIIP was not mentioned as a possible cause of the patient's symptoms. Various terms were used to describe the operative findings, such as superior mesenteric artery syndrome, functional megacolon, paralytic ileus, chronic volvulus, megaduodenum, duodenal aplasia, and partial bowel obstruction.

2) Certain procedures were not clinically helpful. None of the three patients who had gastrojejunostomies and none of the three patients who had small bowel resections improved. In addition, no patient improved after lysis of adhesions or small bowel plications.

3) Certain operations may be beneficial in carefully selected patients. Two patients (cases 1 and 4), who had megaduodenums along with normal stomachs and jejunums, improved after side-to-side duodenojejunostomy. One patient (case 4) had a recurrence of vomiting and abdominal distention 14 years after initial examination. Although initially he had duodenal involvement alone, at the time of his clinical recurrence the entire small intestine was involved as well. Because the patient had persistent vomiting of large amounts of bile stained fluid, the authors believed that the patient might benefit from a procedure which defunctionalized the duodenum as much as possible. The operation consisted of detaching the stomach from the duodenum, draining the duodenum dependently into the jejunum with a Roux-en-Y duodenojejunostomy, and decreasing the volume of the duodenum by internally plicating the anterior duodenal wall. Vagotomy and antrectomy were also performed in order to decrease acid production and to improve gastric emptying of both liquids and solids. The follow-up period is, thus far, too short (three months) to judge the effectiveness of this operation, but the patient has improved significantly to date.

Two patients (cases 2 and 10) failed to improve after duodenojejunostomy. It is possible that the patient in case 10 failed to improve because the duodenojejunostomy did nothing to correct his abnormally slow gastric emptying. A Finney pyloroplasty performed four months later stopped the vomiting, although it is unclear how this procedure might have improved gastric emptying.

One patient (case 2) had small bowel and duodenal involvement, so that creating a simple side-to-side connection between an abnormal duodenum and an abnormal jejunum did nothing to enhance intestinal flow.

One patient (case 3) also had small bowel dilatation in addition to a megaduodenum. Like case 4, she improved after having a Roux-en-Y procedure performed in conjunction with mechanical reduction in the size of her patulous duodenum.

4) Some patients who have predominantly colonic symptoms, such as case 7, may benefit from colonic surgery. In addition to case 7, the patient in case 11 was diagnosed as having an idiopathic megacolon in childhood and had a sigmoid colectomy in adolescence. The patient did well on laxatives until many years later, when her small bowel became involved and abdominal distention and vomiting became her predominant symptoms.

Two patients did not benefit from segmental colonic resections. In both patients (cases 6 and 12) the predominant symptoms were small bowel and no benefit should have been expected.

5) Failure to make a correct diagnosis can lead to clinical confusion and a vicious cycle of repetitive operations. In addition to the 17 operations of case 2, the patient in 12 had 34 operations before a diagnosis of CIIP was made. She currently has a short bowel syndrome and disabling diarrhea, and requires home parenteral nutrition.

Four patients have died, none as a result of surgery. Two patients died as a direct result of CIIP; the patient in case 9 died at the age of 64 years from progressive inanition, and the patient in case 11 died at the age of 35 years of aspiration of gastric contents. The other two patients died of a stroke and of pneumonia, respectively.

We are currently following the clinical courses of three additional patients with CIIP who have not had

TABLE 1. *Surgical Data*

Disorder	Case	Total Operations	Predominant Clinicoanatomic Pattern	Major Procedures Performed in Attempt to Correct Symptoms[1]	Results	Years Since Last Major Operation
Hollow visceral myopathy	1	1	Duodenal & esophageal	Side-to-side duodenojejunostomy	Vomiting ceased; dysphagia persists	15
	2	17	Duodenal & small bowel	Side-to-side duodenojejunostomy; gastrojejunostomy; vagotomy, antrectomy, & Billroth II anastomosis; resection of 50% of small bowel; revision of gastrojejunostomy & Roux-en-Y duodenojejunostomy	Short bowel syndrome; still has abdominal pain & vomiting; requires home parenteral nutrition.	5
	3	3	Duodenal & small bowel	Gastrojejunostomy; revision of gastrojejunostomy, duodenoplasty & Roux-en-Y duodenojejunostomy	Symptoms recurred after gastrojejunostomy, but markedly improved after last operation; no hospitalization since	5
	4	5	Duodenal initially; duodenal — small bowel later	Side-to-side duodenojejunostomy; vagotomy, antrectomy, duodenoplasty, Billroth II gastrojejunostomy & Roux-en-Y duodenojejunostomy	Was asymptomatic for 14 years after duodenojejunostomy; improved after last operation	0.3
Familial visceral neuropathy	5†	1	Small bowel	Gastrojejunostomy	No improvement	8
	6†	1	Esophageal & small bowel	Segmental resection of left colon many years ago for diverticular disease	No improvement	22
Sporadic visceral neuropathy	7	1	Colonic	Subtotal colectomy & ileorectal anastomosis	Improved	0.5
Jejunal diverticulosis	8	4	Small bowel	Resection of 90 cm of small bowel; small bowel plication	No improvement; requires home parenteral nutrition	15
Miscellaneous	9†	2	Small bowel	Repair of incarcerated umbilical hernia; reduction of small & large bowel volvulus	No improvement	1
	10	2	Gastroduodenal	Side-to-side duodenojejunostomy; Finney pyloroplasty	No improvement until Finney pyloroplasty done 4 months later; now doing well	2
	11†	2	Colonic; later small bowel	Resection of sigmoid colon	Constipation improved but still needed laxatives; died of aspiration when small bowel became involved later	
	12	34	Small bowel	Small bowel and colonic resections	Short bowel syndrome; requires home parenteral nutrition	5

* Many operations were exploratory or involved lysis of adhesions; only the major procedures are mentioned in this column.

† Deceased.

surgery. One is the patient with esophageal symptoms reported above. The second patient is improved and maintaining weight while on home parenteral nutrition. The third patient has had several acute attacks, but is currently doing well on a diet restricted in fat and lactose.

Discussion

CIIP is a clinical syndrome caused by a variety of disorders that inhibit intestinal propulsion. It should be diagnosed by obtaining gastrointestinal contrast radiographs and esophageal manometric studies in patients

with suspicious clinical presentations, *i.e.*, recurrent vomiting, abdominal pain and distention, diarrhea and/or constipation, and weight loss. The entire gastrointestinal tract, from esophagus to anus, should be evaluated to determine whether there is widespread evidence of abnormal motor activity and intestinal dilatation,[25] and a pattern consistent with classical or atypical achalasia should be sought in the manometric study.[24] If the diagnosis can be established by finding evidence of widespread abnormal motility in the absence of a mechanically obstructing lesion, an exploratory laporatomy will be unnecessary and, in fact, could be meddlesome. Because the natural history of this disease is characterized by remissions and exacerbations, an acute relapse subsequent to an exploratory laporatomy will raise the fear of true mechanical obstruction from the previous operation, thus prompting a second exploration. A purely diagnostic laparotomy could thereby establish a cycle of repetitive operations. On the other hand, there may not be enough time to establish a diagnosis in a patient presenting for the first time with an acute attack, so that some patients will have exploratory laparotomies during which no mechanical cause for obstruction will be found. CIIP should be considered if one or more segments of the gastrointestinal tract are dilated in the absence of a mechanical cause. Whether or not to biopsy the bowel at exploratory laparotomy is unclear. There is no specific treatment for any of the histologic variants of CIIP while there is some risk of additional complications. However, we currently believe that a biopsy specimen should be obtained, even in the absence of specific medical management, to establish a firm diagnosis, determine the prognosis for the patient, and to plan for follow-up treatment. If a biopsy specimen is obtained, it should be full thickness and approximately 2×2 cm in order to use a special neuropathologic silver stain in addition to the routine histologic tests.[26] In some biopsy specimens the nature of the histologic abnormality will be apparent only with this special technique. We feel that no palliative procedure is indicated at this time and that nothing should be done except for the biopsy. Postoperative esophageal manometric studies and contrast radiographs should be obtained to further strengthen the diagnosis of CIIP and define the pattern of involvement.

The optimal role of surgery should be that of elective palliative treatment of disabling and medically unresponsive symptoms. The major clinical symptoms should be carefully defined and correlated with the radiographic and manometric data to determine whether the patient has predominance of an esophageal, gastroduodenal, small bowel or colonic syndrome. Although each patient may have anatomic or physiologic involvement of the entire gastrointestinal tract and multiple gastrointestinal complaints, one of these clinicoanatomic patterns may predominate. Once a dominant pattern has been determined in the individual patient who remains disabled despite optimal medical management, a specific surgical approach can be recommended. We proceed as follows for each of the clinicoanatomic patterns:

I. *Esophageal.* If dysphagia and food retention within the esophagus are the predominant complaints, and esophageal manometric studies demonstrate a pattern of classical or atypical achalasia, balloon esophageal dilatation should be considered. Failure to respond to balloon dilatation could be an indication for a Heller myotomy, although we have no data on this point as of yet.

II. *Gastroduodenal.* If the primary complaints are abdominal pain and vomiting, and a barium contrast study shows a megaduodenum with or without gastric obstruction, we attempt to defunctionalize the duodenum by dividing it from the stomach and anastomosing it to the jejunum in a Roux-en-Y fashion. Because this is a potentially ulcer producing operaton, a vagotomy and antrectomy are also performed. The objective of this operation is to allow the duodenum to empty by gravity and to shunt the gastric contents past the duodenum. In tremendously distended duodenums a duodenoplasty or removal of a segment of the antimesenteric border of the duodenum is recommended to reduce the duodenum's volume and prevent an excessive accumulation of pancreaticobiliary secretions. Of our five patients who had operations performed for megaduodenum, two had this procedure performed, and both patients symptomatically improved.

The alternative operation of side-to-side duodenojejunostomy could be performed on selected patients who have normal stomachs and jejunums, and whose only abnormality is a megaduodenum. This operation was successful in two patients who fit these criteria. However, one of the two patients had a recurrence of the symptoms 14 years later when the rest of his small intestine became involved.

Table 2 reviews previous reports of patients who had surgery performed for megaduodenum. Most patients had relatively short follow-up periods. However, it is apparent from Table 2 that some patients improved with duodenojejunostomy alone. Unfortunately, the reports do not relate the total extent of intestinal involvement, so that it is not possible to determine whether the patients who

TABLE 2. *Results of Surgery for Megaduodenum*

Reference	Operative Findings	Procedures	Histologic Findings	Results	Period of Follow-up
Barnett et al.[2]	Dilated duodenum	Retrocolic duodenojejunostomy	Ganglion cells replaced by "sheath cells"	Improved	7 months
Matzen et al.[13]	Dilated duodenum	Duodenojejunostomy	Degeneration of ganglion cells	Died postoperatively	
Uncapher et al.[28]	Dilated stomach and duodenum	Duodenoplasty	Normal	Improved	4 years
Nordentoft[19]	Dilated duodenum	Duodenojejunostomy	Normal	Improved	Weeks
Newton[18] case 1	Dilated duodenum	Multiple operations: 1) Division of ligament of Treitz 2) Partial gastrectomy and gastrojejunostomy 3) Side-to-side duodenojejunostomy 4) Vagotomy, duodenal resection & duodenojejunal anastomosis	Normal	Operations 1–3: not improved Operation 4: improved	6 years
case 2	Dilated duodenum	Vagotomy, partial gastrectomy, duodenal resection & duodenojejunal anastomosis	Normal	Improved	3 months
Fischer[6]		Duodenum resected & duodenojejunostomy	Normal	Continued symptoms	1 month
case 2	Dilated duodenum				
case 3	Dilated duodenum	Duodenojejunostomy		—	none
Law et al.[8]	Dilated duodenum	Duodenojejunostomy	Normal	Improved	2 years
Anuras et al.[1]					
case 4	Dilated duodenum	Duodenojejunostomy	Familial visceral myopathy (FVM)	Died postoperatively	
case 6	Dilated stomach and duodenum	Gastrojejunostomy	FVM	Not improved	
case 12	Dilated duodenum	Six operations including duodenojejunostomy	FVM	No long-term improvement	
case 15	Dilated duodenum	Side-to-side duodenojejunostomy	FVM	Improved	6 years
case 16	Dilated duodenum	Side-to-side duodenojejunostomy	FVM	Improved	1½ years
Weiss[29]					
case 1	Dilated duodenum	Duodenojejunostomy	—	Improved	7 years
case 2	Dilated stomach, duodenum and proximal jejunum	Partial gastrectomy with gastrojejunostomy	—	Not improved; died 1 yr later	
case 3	Dilated duodenum	Duodenojejunostomy	—	Died 17 days postoperatively	

improved had involvement limited to the duodenum. Some patients also improved after duodenoplasty or duodenal resections, although in these cases, too, the extent of intestinal involvement was not usually reported. We agree with Anuras et al. that antibiotics should be administered before, during and after surgery to decrease the chance of postoperative complications caused by bacteria-rich duodenal fluid.[1]

III. *Small bowel.* Surgery has not been helpful and indeed has often been meddlesome in patients whose predominant complaints were marked abdominal distension and pain associated with diffuse dilatation of the small intestine. We feel that exploratory laporatomy is not indicated in a patient whose predominant complaint concerns the small bowel, and that it may only serve to initiate a cycle of repetitive operative procedures.

There is nothing in the literature to support any other medical course (Table 3). Only two of the 12 reported patients clinically improved, but the follow-up period was only four months in one patient, while the other patient developed symptoms of "subacute intestinal obstruction" within ten months of surgery.[3,20]

IV. *Colonic* Because of the colon's inability to have organized propulsive contractions, the colon in CIIP acts as a functional obstruction to gas and stool. The procedure we recommend in patients incapacitated by this pattern of involvement is total abdominal colectomy with ileal anastomosis performed at the peritoneal reflection. This recom-

TABLE 3. *Results of Surgery for Small Bowel Involvement*

Reference	Operative Findings	Procedures	Histologic Findings	Results
McClelland et al.[11]	Dilated small bowel	Two exploratory laparotomies with small bowel plication	Normal	Patient died without relief
Naish et al.[17]	Dilated small bowel	Two exploratory laparotomies; appendectomy	Vacuolated muscle	Not improved
Murley[15]	Dilated small bowel & duodenum	Appendectomy and biopsy of ileum	Degenerated muscle	Mildly improved
Nahai[16]	Dilated small bowel	Two exploratory laparotomies with biopsy & Baker tube insertion	Hypertrophied muscle	No long-term follow-up period
Maldonado et al.[12] (five patients)	Four patients with variable amounts of dilated small bowel; 1 with dilated colon & small bowel	Multiple types of procedures See table in reference 12	No significant abnormalities	None given
Dyer et al.[3]	Dilated small bowel & duodenum	Multiple laparotomies; vagotomy & gastrojejunostomy; resection of part of duodenum & 100 cm jejunum; antrectomy & Roux-en-Y duodenojejunostomy	Degenerated myenteric plexus	Mild complaints persisted
Paul et al.[20]	Dilated small bowel from duodenum to within 4 ft of ileocecal valve	1) Exploratory laparotomy; 2) resection of small bowel from Treitz to 230 cm proximal to ileocecal valve	None	Improved, but only four-month follow-up period
Anuras et al.[1]	Dilated distal jejunum & proximal ileum (case 10)	1) Exploratory laparotomy 2) Lysis of adhesions 3) Lysis of adhesions 4) Stamm jejunostomy & lysis of adhesions	Familial visceral myopathy	Not improved until last operation; improved over 12 years of followup

mendation is not made wholly on the basis of our own experience or on the very limited experience reported with CIIP, but rather it is made on the recomendation that is reported in the literature on idiopathic megacolon. In the two largest series reported, total abdominal colectomy gave the best results and had a low morbidity rate.[7,10] By anastomosing the ileum in the peritoneal cavity, the operative morbidity rate should be reduced, while keeping the colonic remnant at a minimal length. By resecting all of the abdominal colon the risk of volvulus is remote and the chances of recurrent colonic symptoms are reduced.

A word of caution is in order because patients with CIIP do not have involvement of the colon alone, as do the patients with idiopathic megacolon. In patients with CIIP, involvement of the small intestine may lead to bacterial overgrowth with subsequent malabsorption and diarrhea at any time during the duration of this illness. Diarrhea could prove to be particularly troublesome in a patient in whom colectomy and ileorectal anastomosis were performed. Careful consideration should be given to this potentially complicating factor before the decision to perform a colectomy is made.

We emphasize that these recommendations are made on the basis of limited data. However, patients with CIIP are being increasingly recognized at a time when little is known about what should be done to ameliorate their symptoms. A carefully chosen operative procedure may be one possible answer. Patients who have procedures for defined clinicoanatomic patterns should be thoroughly studied beforehand, have regular follow-up examinations, and reported in the literature. Only with more data will we know whether any operative procedure will help to improve the quality of life of these patients.

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