46 Chronic Intestinal Pseudo-obstruction Syndrome: Surgical Approach and Intestinal Transplantation

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Chronic intestinal pseudo-obstruction syndrome (CIPO) is a severe, often unrecognized cause of neonatal or post-natal progressive intestinal failure (IF). This rare syndrome represents one of the main causes of IF and is characterized by impairment of physical growth and development as well as by a high rate of morbidity and mortality.

The diagnosis of CIPO is based on typical clinical manifestations, radiological evidence of distended bowel loops with air-fluid levels, and the exclusion of any organic obstruction of the gut lumen [1–5]. CIPO is often unrecognized, and the diagnosis, therefore, delayed by several years with useless and potentially dangerous surgeries.

CIPO can occur in patients with underlying diseases associated with gastrointestinal manifestations (scleroderma, amyloidosis, hypothyroidism, etc.) or be secondary to water-electrolyte disorders (e.g., hypokalemia), and toxic, viral, and parasitic causes. Most cases are idiopathic and sporadic, even though familial forms with either dominant or recessive autosomal inheritance have been described. Based on histological features intestinal pseudo-obstruction is classified into three main groups: neuropathies, and myopathies or "mesenchymopathies," according to the predominant involvement of enteric neurones, smooth muscle cells, and interstitial cells of Cajal, respectively [6–14]. Mitochondrial disorders have been reported [15, 16]. Regardless of the histologic type, CIPO always involves alterations of smooth muscle contractile function, leading to abnormal intestinal tract peristalsis and nutritional disorders. Manometry can play a supportive role in defining the diagnosis, as well as by showing differences in the manometric pattern of CIPO [17]. Accompanying uropathies must be sought in patients with CIPO [6, 18]. The clinical impact of these uropathies may be important and require specific management by using daily drainage and, sometimes, vesicostomy.

Longitudinal surveys have been published, including a large multicenter French pediatric study [19–23]. Long-term outcomes are generally poor despite surgical and medical therapies and characterized by disabling and potentially life-threatening complications. Treatment of CIPO involves nutritional, pharmacological, and surgical therapies but is often frustrating and does not change the natural course in the majority of cases [24–27]. Nutritional management is of crucial importance in the pediatric age group and involves enteral delivery of special formulae, by nasogastric tube, percutaneous gastrostomy, or

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jejunostomy [26]. In the most severe cases, parenteral nutrition becomes mandatory in order to satisfy nutritional requirements and appropriately manage obstructive episodes.

Surgery is one of the mainstays of CIPO therapeutic management. Surgery is performed in a variety of situations in pediatric patients but surgical options must be evaluated carefully. There is no consensus regarding indications and procedures. This short chapter aims to review the main situations in which surgery may be required.

Surgery for Diagnosis

Variable clinical presentation and lack of other specific diagnostic tests often leads to surgery being required for diagnosis. Nevertheless, unnecessary laparotomy could be avoided since diagnosis is mostly based on clinical and radiological symptoms of intestinal obstruction. It is not unusual, however, for some patients, especially children and adolescents with an acute presentation, to undergo an exploratory laparotomy. In the absence of organic obstruction observed at this laparotomy, we suggest that a medico-surgical discussion be undertaken to consider:

- Performing intestinal full-thickness biopsies at different levels for histopathologic analysis
- Performing an enterostomy according to the level of intestinal distension

In reality, in most cases, the acute presentation and subsequent surgical procedure do not occur at a specialized center, and these suggested interventions are not done. Such issues are controversial but we do propose that if the diagnosis of CIPO is strongly suggested from the surgical exploration, careful biopsies should be performed. Regarding enterostomy, our experience tends to suggest that if it is not performed at first laparotomy, it will need to be done later but with subsequent increased risk of peritoneal adhesions.

In summary, patients with evidence of CIPO from clinical and radiological presentation should not be operated on to make the diagnosis. Patients who undergo laparotomy for enterostomy because of permanent or recurrent intestinal obstruction should have intestinal full-thickness biopsies for specific diagnosis. This should be done regardless of the patient's age.

Gastrostomy Tubing

Bowel decompression by a gastrostomy and/or jejunostomy is often required. Repeated acute episodes of bowel obstruction as well as chronic intestinal distension require bowel decompression by using nasogastric suction. The placement of a venting gastrostomy is of great benefit in avoiding the recurrent placement of nasogastric tubes. When surgery is required, a gastrostomy may be performed during the same surgical procedure. If a gastrostomy is not surgically placed, percutaneous endoscopic gastrostomy tube (GT) placement is easily achieved in these children. Since enteral feeding should always be preferred to using parenteral nutrition (PN), intragastric administration of feeding may be achieved by the GT as continuous or bolus enteral tube feeding.

Enterostomy

In neonates and young infants, intestinal obstruction may last several weeks requiring total parenteral nutrition (TPN) with subsequent complications including catheter-related sepsis and liver disease [4]. Enterostomy may offer the chance to restart intestinal transit allowing feeding and reducing the need for PN.

In some patients, attacks of intestinal obstruction are frequent and/or life threatening. Chronic bowel dilatation impairs intestinal motility creating a vicious circle which increases intraluminal bacterial overgrowth with the subsequent risk of intestinal translocation, enterotoxin release, and liver disease [28, 29]. Enterostomy should be performed to bypass the functional obstruction and obtain digestive decompression.

The location of the enterostomy is a matter of debate. In cases of obvious megacystis microcolon syndrome, a terminal ileostomy is certainly required. Otherwise, we do recommend performing a terminal ileostomy and avoiding a colostomy

whatever the clinical presentation or histopathologic pattern. It is important to consider the so-called ileo-caecal brake as the segment that should be short-circuited. In our experience, all patients who first underwent a colostomy went on to have formation of a terminal ileostomy or jejunostomy.

The outcome after ileostomy or jejunostomy varies according to the location of the enterostomy and to the disease itself. The literature does not provide any evidence of a histopathology-related prognosis even if the survey reported by Henyeke et al. suggested worse prognosis of myopathies and that they all need ileostomies [22]. However, much fewer than 50% of patients improve after ileostomy by being weaned from PN. In our opinion, enterostomy, as distal as possible is the most logical approach. Terminal ileostomy usually enables transit to resume and leads to a major long-term reduction in obstructive episodes. We currently perform an ileostomy to obtain durable intestinal autonomy and PN weaning, with the future plan to do a total or subtotal colectomy with ileorectal or ileosigmoid pull-through [23].

Irtan et al. have reported stomal prolapse in children with chronic intestinal pseudo-obstruction as a frequent complication [30]. Twenty-two out of 34 (65%) CIPO children referred to their center between 1988 and 2008 had a stoma and were compared with 22 other children referred for another pathology necessitating a stoma. The incidence of stomal prolapse in CIPO children was 45% vs. 9% in non-CIPO children ($p=0.01$). Prolapse occurred between the first postoperative day and the tenth postoperative month, with a median of 2 months. Surgical management was required in 60%, with an intestinal necrosis rate of 20% leading to intestinal resection. The authors did not identify particular risk factors favoring stomal prolapse.

Percutaneous endoscopic cecostomy or colostomy (PEC) is increasingly proposed as an alternative to surgery to treat CIPO and relapsing sigmoid volvulus [31–34]. Cecostomies or even sigmoidostomies have been used to administer antegrade enemas when intractable constipation appears to be the prominent symptom. A few reports are available both in children and adults describing the indications, complications, and outcomes. A retrospective, single-center study involving eight adults was reported by Lynch et al. [33]. Six patients had CIPO and two had chronic constipation. Use in seven of eight cases resulted in clinical improvement with reduction of intestinal obstruction episodes and improved feed tolerance. One patient suffering chronic constipation required surgical removal of the percutaneous endoscopic cecostomy tube at 4 days for fecal spillage resulting in peritonitis despite successful tube placement. Removal of the cecostomy tube occurred in three of six cases of pseudo-obstruction (the other three remain in place). In the other patient with chronic constipation, clinical improvement occurred, but the patient died of underlying illness 21 days after placement. A case of acute stercoral peritonitis was reported [34]. At laparotomy, the colostomy flange was embedded in the abdominal wall but no pressure necrosis was found at the level of the colonic wall. This complication was likely related to inadvertent traction of the colostomy tube. Percutaneous endoscopic cecostomy is considered by some authors as a viable alternative to surgically or fluoroscopically placed cecostomy in a select group of patients with recurrent colonic pseudo-obstruction or chronic intractable constipation.

Closure of the Stoma

In children whom a decompression ileostomy has produced relief, but there is diffuse disease, the urge to re-establish connection with the defunctioned limb of the bowel should be resisted as this will only result in further episodes of obstruction. In other words, performing an ileostomy and closing it because of clinical improvement results in the patient undergoing two surgical procedures without resolution of the primary issues. This should be avoided. Conversely, in patients in which clear improvement from ileostomy is observed, with PN weaning and at least 2 years follow up on enteral tube feeding or oral feeding without exacerbations, total colectomy and

ileorectal anastomosis with the Duhamel procedure may be considered. In our experience, two-third of the patients who underwent this procedure remain off PN for a long period of time [23].

Recurrent Laparotomies and Enterectomy

In the past, many patients underwent multiple surgical procedures. Unnecessary abdominal surgery in children with CIPO should be avoided because they bear the risk of prolonged postoperative ileus and developing adhesions, creating a diagnostic problem each time there is a new obstructive episode. Mechanical obstruction should be considered in patients with an enterostomy who continue to present with exacerbations of bowel obstruction. In an earlier study involving only seven patients, surgery was performed as a treatment 21 times with a mean of three procedures per patient [20]. This is similar to other data reported. In one study, 67 surgical procedures were performed in 22 patients [8], and in another study involving 105 pediatric infants and children, 71 patients underwent surgery during their illness, with 217 surgical procedures [21]. An ostomy was the most performed procedure. Surgery may cause adhesions, so interpretations of postoperative obstructive episodes are difficult. Exploratory laparotomy for obstruction should be performed only when a clear mechanical obstruction has been demonstrated which remains very difficult to assess. Signs of peritonitis, extreme dilatation and pain in association with specific episodes of obstruction point more towards mechanical rather than functional obstruction, and a laparotomy may be required to relieve it.

Patients with CIPO or chronic intractable constipation (CIC) may develop anatomical obstruction such as colonic volvulus, with presenting symptoms mimicking those of underlying pseudo-obstruction. Patient records of 8 children with colonic volvulus were retrospectively reviewed [35]. The mean age at presentation with colonic volvulus was 13.2 ± 5.05 years. All patients presented with worsening of abdominal distension and pain. The mean duration of symptoms of colonic volvulus before seeking medical help was 4.2 days (range 1–7 days). Water-soluble contrast enema was the single most useful investigation for confirming the diagnosis. All patients required surgery. There was no mortality associated with colonic volvulus. Clinicians should be vigilant and include volvulus in the differential diagnosis of the acute onset of abdominal distension and pain in patients with CIPO and CIC. Delay in diagnosis can result in bowel ischemia and perforation.

Some patients, in whom there is segmental bowel dilatation but no evidence of mechanical obstruction, have been reported to benefit from segmental resections or to have improved following placement of a jejunostomy tube within the dilated loop [36, 37]. In our experience, the use of this jejunostomy button device for daily intermittent bowel decompression can effectively improve bowel function allowing decreased PN intake. However, one should consider the quality of life (QOL) of a child with three tubes and, for most of the time, a central line.

Patients suffering from CIPO clearly benefit from home parenteral nutrition (HPN) to maintain adequate nutritional status and general health [38]. However, permanent and severe intestinal dysmotility can seriously disturb the QOL to the point of making it intolerable. Subtotal enterectomy [39, 40] or bilateral thoracoscopic splanchnicectomy have been proposed in severe CIPO [41]. A retrospective study of eight patients with end-stage CIPO maintained on HPN and suffering from chronic occlusive symptoms refractory to medical treatment underwent extensive small bowel resection preserving less than 70 cm of total small bowel and less than 20 cm of ileum [40]. The jejunum was anastomosed either to the ileum or to the colon. Six patients were completely relieved from obstructive symptoms. Two patients needed a second operation to remove the residual ileum because of recurrent symptoms. Both were significantly improved and there was no postoperative death. All patients experienced a significant improvement in their QOL. Near total small bowel resection appears to be a safe and effective procedure

in end-stage CIPO patients, refractory to optimal medical treatment.

The implantation of gastric or intestinal pacemakers aimed at improving motility constitutes a promising investigational approach in patients with severe motility disorders. The use of gastric electrical stimulation has been shown to significantly improve nausea and vomiting not only in patients with diabetic gastroparesis, but more recently also in three adult patients with familial and one with postsurgical CIPO with disabling nausea and vomiting [42]. The weekly vomiting frequency decreased from 24 before implantation of the gastric pacemaker to 6.9 after 12 months. The clinical response was unrelated to the presence of, or improvement in, delayed gastric emptying in these patients. Although placements of the electrodes along the anterolateral surface of the stomach was successful in most patients by laparoscopic implantation, the procedure was not without risk since the electrodes caused ileus necessitating explantation and short intestinal resection [42].

Intestinal Transplantation

Intestinal transplantation (ITx) has become a lifesaving procedure for patients with irreversible intestinal failure (IF) [43–45]. Indications for ITx include not only extreme short bowel syndromes but also all situations in which the small intestine is unable to achieve nutritional requirements; these include inborn errors of intestinal mucosa development (intestinal epithelial dysplasia, microvillus inclusion disease) or severe motility disorders such as CIPO [46–50]. Approved indications for ITx include liver dysfunction, loss of major venous access, frequent central line-related sepsis, and recurrent episodes of severe dehydration despite intravenous fluid management. Surgical options include transplantation of the isolated intestine, combined liver–intestine transplantation, or multivisceral transplantation of the stomach, duodenum, pancreas, and small bowel (with or without the liver). Immunosuppression for ITx is based on tacrolimus therapy, often with induction immunosuppression using antilymphocyte antibodies (e.g., antithymocyte antibody and alemtuzumab). Experience at centers of excellence demonstrate 1- and 5-year patient survival rates of 95% and 77%, respectively, with ongoing investigations focusing on lowering long-term causes of graft loss such as chronic rejection [45].

In many cases of CIPO, outcome is poor, with a constant risk of sepsis from intestinal bacterial overgrowth, and water-electrolytic disorders related to intraluminal fluid retention. ITx is the only definitive curative treatment especially when many medical and surgical attempts have failed. ITx with or without liver transplantation is required in patients with primary neuro-muscular disease and PN-related complications such as progressive or end-stage liver disease or for those whose intravenous access has become unreliable and precarious because of repeated sepsis and extensive thrombosis. Transplant procedures vary according to indication for liver transplant and based on the experience of the transplant surgical team [47–49]. Combined small bowel–liver transplantations or multivisceral transplantations including the stomach have been performed in refractory forms of CIPO associated with end-stage liver disease [47–49]. Multivisceral transplantation (MVTx) was reported in 16 children with a median age of 4 years [47]. Indications for MVTx were liver failure ($n=10$), loss of venous access ($n=3$), or sepsis ($n=3$). Modified MVTx without the liver was performed in six patients. Reported actuarial patient survival for 1 year/2 years were 57.1% to 88.9%/42.9% to 77.8% according to immunosuppressive regimens. Currently, none of the long-term survivors are on PN and all tolerate enteral feeding. Gastric emptying was substantially affected in one case. Bladder function did not improve in those with urinary retention problems. MVTx for CIPO offers a lifesaving option with excellent function of the transplanted pancreas and stomach among survivors.

ITx may represent the only definitive cure for patients with permanent intestinal failure due to CIPO. However, graft rejection, and immunosuppression-related lymphoproliferative disorders are more common than other organ transplants. It is not yet established whether the results of ITx achieved in CIPO patients are

equivalent to those experienced with other causes of IF such as short gut syndrome, total aganglionosis, microvillous inclusion disease, or epithelial dysplasia [51]. Complications seem to be more common due to multiple previous abdominal surgeries, dysmotility of the stomach and esophagus, and extra-intestinal manifestations including associated anomalies of the urological, immune, and neurological systems. An extensive workup including a search for mitochondrial disorders should be performed before any discussion of ITx and careful consideration is required before transplantation is undertaken. Determining the extent of the disease process (which may involve any part of the gastrointestinal tract) and the type of organ transplantation required is mandatory. Early referral is essential on initial presentation of these patients to enable optimal medical care and ensure that transplantation remains an option [43, 52].

Ethical dilemmas may arise with children who will never be able to tolerate full enteral feeding. Some patients with severe CIPO may be disabled because of chronic, massive GI dilatation refractory to stomal decompression or partial enterectomy. The poor QOL might serve as an indication for ITx, and not the usual criteria, which include progressive liver disease, loss of vascular access, and repeated life-threatening sepsis. In any case, parents must be extensively informed about the risks of the procedure and about the outcomes of all decisions.

Conclusion

Primary CIPO is a rare condition with a variable clinical expression. Medical management remains difficult and prognosis poor. Histological studies are essential to classify the syndrome, even if manometric data are able to differentiate between myopathic and neuropathic forms, and although histological type does not appear to influence management and long-term outcome. A trained multidisciplinary team, including surgeons, gastroenterologists, nutritionists, and a home PN coordinator, should assume the management of these patients which may involve a PN program and transplant surgery. For many reasons (nutrition, prevention of infectious complications, etc.), an enterostomy (preferably an ileostomy) is often performed as one of the first therapeutic measures. The "permanent" surgical reconstruction, designed to be minimally obstructive, is only envisaged after a long period of stability and if possible when the child is weaned from long-term PN. Intestinal transplantation may be the last therapeutic option when all medical and surgical approaches have failed.

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