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Chronic intestinal pseudo-obstruction in adult patients: multidetector row helical CT features

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Abstract Chronic intestinal pseudo-obstruction (CIPO) is a rare condition due to severe gastrointestinal motility disorder. Adult patients with CIPO experience symptoms of mechanical obstruction, but reliable clinical signs that may help distinguish between actual mechanical obstruction and CIPO are lacking. Additionally, abdominal plain films that commonly show bowel dilatation with air-fluid levels do not reach acceptable degrees of specificity to exclude actual obstruction. Therefore, most adult patients with CIPO usually undergo multiple and often fruitless surgery, often leading to repeated bowel resections before diagnosis is made. In these patients who present with abdominal signs mimicking symptoms that would warrant surgical exploration, multidetector-row helical CT (MDCT) is helpful to resolve this diagnostic dilemma. MDCT shows a diffusely distended bowel and helps to rule out a mechanical cause of obstruction, thus suggesting CIPO and obviating the need for unnecessary laparotomy. In adult patients with CIPO, MDCT may show pneumatosis intestinalis, pneumoperitoneum or intussusception. However, these conditions generally do not require surgery in patients with CIPO. This pictorial essay presents the more and less common MDCT features of CIPO in adult patients, to make the reader more familiar with this disease.

Keywords Abdominal imaging–abdomen · CT–computed tomography-chronic intestinal pseudo-obstruction–intestine disease-chronic intestinal pseudo-obstruction

Introduction

Intestinal pseudo-obstruction refers to a syndrome that mimics bowel obstruction, with failure of intestinal motility in conjunction with abdominal pain and intestinal distension, but in the absence of a mechanical cause. This situation may be observed acutely in hospitalized patients with serious underlying medical or surgical conditions [1]. Acute intestinal pseudo-obstruction corresponds to a paralytic ileus, is characterized by the lack of motor activity and is a self-limited condition. The most common cause of acute intestinal pseudo-obstruction, also known as Ogilvie's syndrome, is a transient postoperative ileus [2]. Typically, there is an acute and marked colonic distension, which can be managed by colonic decompression or with an anticholinesterase such as neostigmine [3]. In contrast, chronic intestinal pseudo-obstruction (CIPO) is a rare clinical condition due to a severe gastrointestinal motility disorder. CIPO is clinically characterized by repetitive or chronic symptoms of bowel obstruction in the absence of a fixed, lumen-obstructing cause or metabolic disorders [4, 5]. Adult patients with CIPO experience symptoms of mechanical obstruction (including abdominal pain, bowel distension, nausea and vomiting), but reliable clinical features that may help distinguish between actual mechanical intestinal obstruction and CIPO are lacking. In addition, abdominal plain films, which commonly show evidence of bowel dilatation with air-fluid levels, do not reach acceptable degrees of specificity to exclude actual bowel obstruction. For these reasons, a majority of adult

patients with CIPO usually undergo multiple and fruitless abdominal explorations, often leading to repeated bowel resections before diagnosis of the disease is made [6]. In our institution, adult patients with CIPO are investigated with multidetector-row helical CT (MDCT) imaging, which proved to be more effective than abdominal plain films and follow-through studies in the diagnosis of acute small bowel obstruction [7]. Once the diagnosis is made, acute episodes of exacerbation of the chronic symptoms may lead to exploratory laparotomies. In such situations, we found that MDCT is a helpful imaging modality for multiple reasons. MDCT imaging is performed either to rule out a potential curable cause of bowel obstruction or to depict intra- or extraintestinal tract complications.

The goal of this pictorial essay was to present the more and the less common MDCT features of CIPO in adult patients, with the aim of making the reader more familiar with this disease for which the majority of patients undergo multiple and potentially dangerous abdominal surgeries [6].

Clinical background

Pathophysiologically, CIPO is characterized by ineffective intestinal propulsion due to an intestinal smooth muscle fiber deficiency and/or an intrinsic enteric nervous system disease. In adult patients, CIPO can be paraneoplastic, can be induced by drug therapy or may be secondary to a variety of diseases, including scleroderma, amyloidosis, systemic lupus erythematosus, dermatomyositis, rheumatoid arthritis, progressive systemic sclerosis, dermatomyositis, Sjogren's syndrome and diabetes. More rarely, CIPO can be primary (the so-called "idiopathic form") with either a neurogenic or a myogenic origin [4, 6]. The neurogenic form involves the extrinsic innervation of the gastrointestinal tract or intrinsic nerves and intersticial cells of Cajal [8]. The myogenic form involves the intestinal musculature [4, 6]. Familial forms as well as sporadic idiopathic forms of CIPO are well documented [9].

Gastrointestinal manometric studies are used to exclude scleroderma and to differentiate between myogenic and neurogenic forms. A neurogenic origin is suggested when contractions have normal amplitude, but are incoordinated with an abnormal propagation. A myogenic origin is suggested by a diffusely reduced amplitude with a normal propagation [8, 10]. As a limitation, in case of severe, long-standing disease with marked gastrointestinal dilatation, the results of manometric studies may be nonspecific, with only a few contractile activity detected [9]. For this reason, manometric results are considered neither specific nor sensitive enough to serve as accurate markers of the disease by some authors [11, 12]. For others, gastrointestinal manometry carries some degrees of importance because it provides information relative to the status of the different segments of the gastrointestinal tract.

Histopathological diagnosis requires immunofluorescence staining or immunohistochemistry techniques, which are not always available in a routine setting. Also, laparoscopic full-thickness small bowel and colonic biopsies are needed to determine the form of the disease [13]. In patients with a myogenic form of CIPO, histological examination shows thinning and degeneration of the smooth tissue muscle with replacement by fibrous tissue [8]. In patients with a neurogenic form, histological study shows abnormalities in argyrophilic neurons and cellular infiltration of the myenteric plexus. It has been demonstrated that 58% of patients with idiopathic CIPO have a complete or partial deficiency of alpha-actin epitope staining in the inner circular layer of small bowel smooth muscle. This deficiency is considered an important biomarker of idiopathic CIPO [13]. Also, C-kit receptors' staining shows abnormalities in the distribution and number of intersticial cells of Cajal in idiopathic CIPO [14].

Treatment options

Management of adult patients with CIPO involves multiple modalities [15]. Nutritional support, either enteral or parenteral, is often needed because of weight loss and severe malnutrition. It can be done using jejunal feeding through a jejunostomy tube or using a short-term or long-term total parenteral nutrition. Adult patients with CIPO are often intolerant to enteral nutrition, and usually they become dependent on parenteral nutrition. Pharmacologic treatments are used with the aim to improve gastrointestinal motility and contraction of the gastrointestinal tract. They include prokinetic agents (such as metoclopramide and erythromycin) and serotonin receptor agonists (such as cisapride and octreotide) [16–18]. Isolated small bowel transplantation can be proposed in a subset of patients who do not tolerate parenteral nutrition [19]. For patients with a known CIPO presenting with acute exacerbation of the chronic symptoms, laparotomy should not be considered when abdominal pain is minimal and when peritoneal signs, fever and leukocytosis are absent.

Conventional imaging features

Abdominal plain radiographs often show bowel dilatation with multiple air-fluid levels, thus suggesting bowel obstruction (Fig. 1) [20]. In some cases, a marked distension may also involve the stomach and the duodenum. Follow-through studies can be performed to exclude a fixed lumen-occluding lesion, but they are obtainable in a subset of patients with limited distension who can tolerate oral administration of contrast material. In addition, follow-through studies are contributive only when the whole small bowel is filled up to the ileocecal junction, and as a

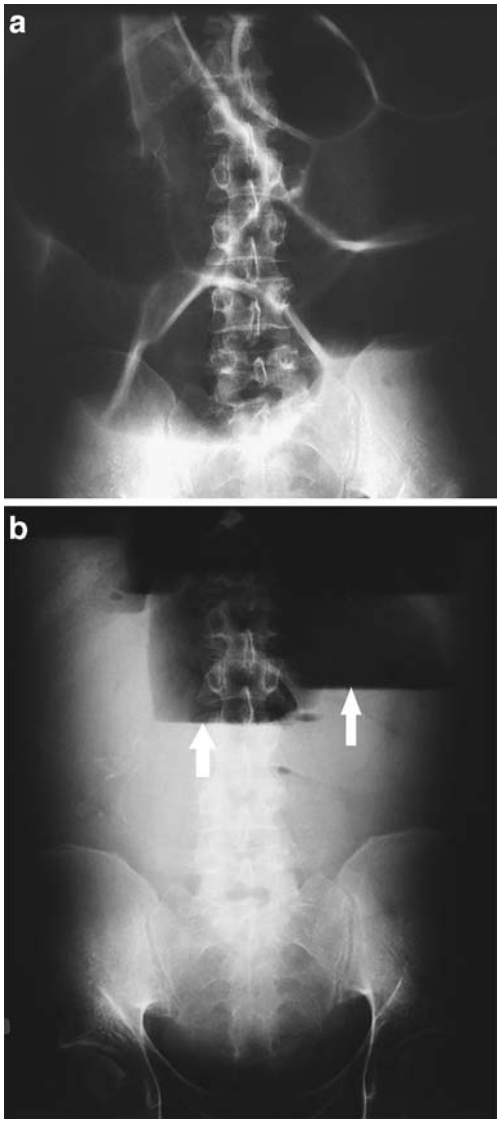

Fig. 1 A 19-year-old man with a known myogenic form of CIPO and prior history of colectomy followed by distal ileostomy. The patient was admitted for routine nutritional evaluation. **a** Abdominal plain radiograph performed in supine position shows multiple distended bowel loops. **b** Abdominal plain radiograph performed in upright position shows air-fluid levels (arrows) within distended bowel loops

limitation flocculation of barium within small bowel distended loops is frequently observed (Fig. 2). Although a water-soluble contrast meal may be used by clinicians to rule out obstruction as an alternative to a small bowel barium study [10], in our institution, upper series and small bowel follow-through examinations are no longer performed. We found both studies time consuming and poorly informative in the majority of cases because of very slow transit time and dilution of the contrast agent. In contrast, MDCT provides readily pertinent and useful information regarding the caliber of the gastrointestinal tract and the status of the bowel wall, thus contributing to prompt and appropriate patient management.

MDCT protocol

MDCT is performed from the dome of the diaphragm through the symphysis pubis. Oral contrast agent is not necessary because of the spontaneous bowel distension due to the disease. Contrast enema is of limited value in the majority of cases, but can be used in selected cases to better image an area of collapsed colon. Because of the clinical presentation, non-ionic iodinated contrast material is administered intravenously at a dose of 120 ml and a rate of 3 ml per second to exclude any possible mesenteric ischemia that might be responsible for the clinical symptoms. Imaging is ideally performed 50 s after the start of bolus injection, when bowel wall enhancement is optimal (enteric phase) [21].

The CT parameters are 120 kVp; 220 mA; 4×2.5 mm detector configuration. The axial data are reconstructed twice, first with 5-mm thickness at 5-mm intervals and then with a 2.5-mm thickness at 2-mm intervals. The second set of reconstructed axial images is then reformatted in the coronal plane with a thickness of 3 mm at 3-mm intervals [22]. Reconstructed images are rapidly obtained on a console system. Although MDCT images obtained in the axial plane provide pertinent information in the majority of cases, adjunct of coronal plane images may improve the reader's confidence level in excluding actual bowel obstruction.

Usual findings

In patients referred for bowel distension, MDCT is an important diagnostic tool for suggesting the diagnosis of CIPO when no causes of obstruction are identified on MDCT images. In these patients who present with abdominal signs that mimic symptoms that would warrant surgical exploration, MDCT imaging is of great help to resolve this diagnostic dilemma. In many cases, MDCT shows a diffusely distended bowel and helps to rule out a mechanical cause of bowel obstruction, thus suggesting CIPO and obviating the need for unnecessary laparotomy. In symptomatic patients with known CIPO, MDCT images provide helpful information to determine the extent and severity of the disease and to depict any possible associated abdominal diseases (Figs. 3, 4 and 5). The degree of dilatation may vary with time or be similar on consecutive MDCT examinations. It has been suggested that patients with a myogenic form might have more frequently marked dilatation of the gastrointestinal tract compared to patients with a neurogenic form [11]. Dilatation of the third portion of the duodenum is commonly observed in adult patients with CIPO secondary to scleroderma, but can be observed

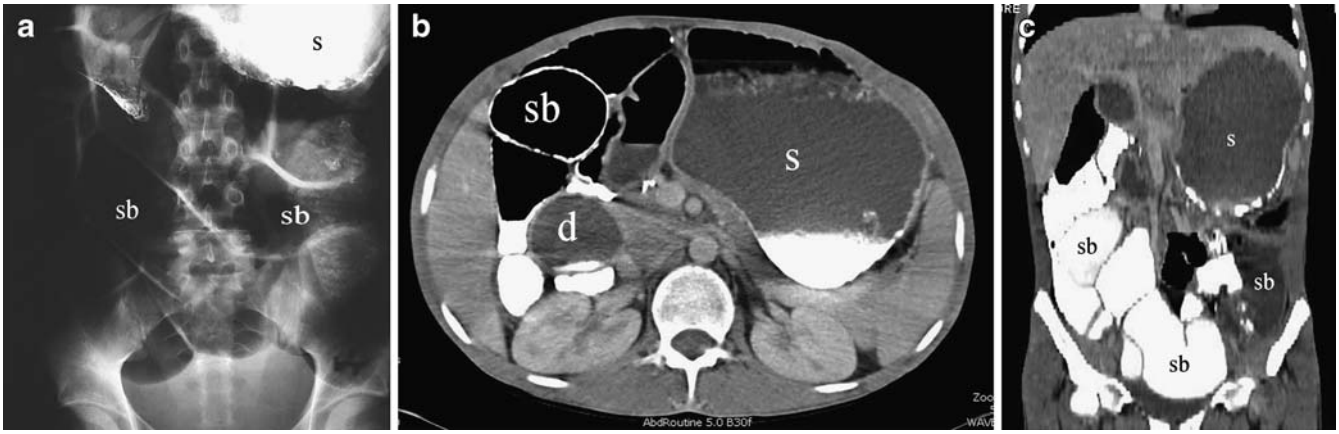

Fig. 2 A 17-year-old man with CIPO secondary to heterozygotic porphyria and prior history of multiple laparotomies and total colectomy followed by ileorectal anastomosis. The patient complained of vomiting. **a** Plain radiograph performed 4 h after oral administration of barium shows a markedly distended stomach (s) filled with barium and multiple air-fluid levels within distended small bowel loops (sb), which does not allow excluding a mechanical cause for bowel obstruction. Follow-through study was inconclusive because of barium stasis in the stomach. **b** Axial MDCT shows markedly distended stomach (s) and duodenum (d) and distended small bowel loops (sb). **c** Coronal CT reformation of MDCT shows marked dilatation of the stomach (s) and entire small bowel. No mechanical cause of obstruction is seen. No surgery was performed, and the patient was managed with conservative therapy

in any adult patient with another cause of CIPO. The small bowel is the most frequently distended portion of the gastrointestinal tract (77%), followed by the duodenum (71%), the stomach (30%) and the colon (22%) [23]. High degrees of distention may prompt the surgeon to operate in case of failure of conservative therapies, depending on the clinical status and the results of biological tests.

It is commonly admitted that in patients with small bowel distension and air-fluid levels, a change in bowel caliber is indicative of mechanical obstruction [7]. However, in adult patients with CIPO, such changes may be observed in the absence of a mechanical obstruction (Fig. 3). As a consequence, a change in small bowel caliber may make exclusion of adhesion difficult.

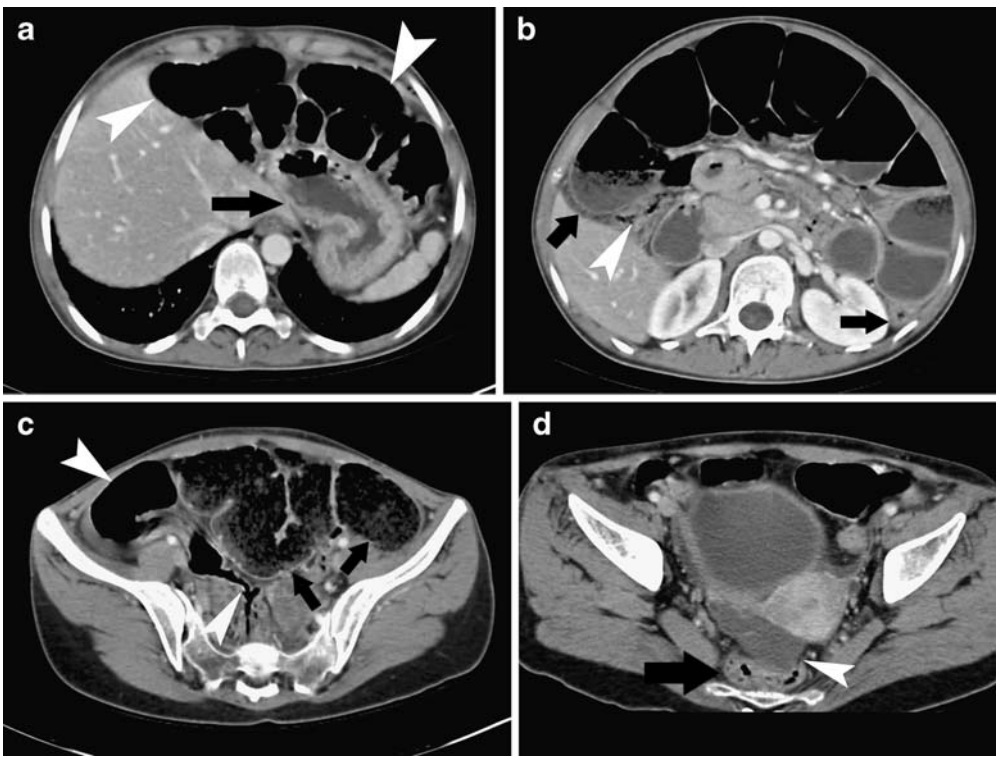

Fig. 3 A 22-year-old woman with a known myogenic form of CIPO. The patient presented with abdominal pain and vomiting. Axial MDCT images obtained at different levels of slice show marked distension, which is limited to the small bowel. No mechanical cause of obstruction is seen. No surgery was performed, and the patient was managed with conservative therapy. **a** At this level of slice, distension involves the small bowel (arrowheads), but not the stomach (arrow). **b** Distension does not involve the ileo-caecal junction (arrowhead), which is flat. The colon (arrows) is collapsed. **c** At this level of slice, the sigmoid colon is empty and not distended (arrowheads). The small bowel is distended and filled with feces (arrows). **d** The rectum is empty (arrow). A small amount of free fluid is seen in the Douglas pouch (arrowhead)

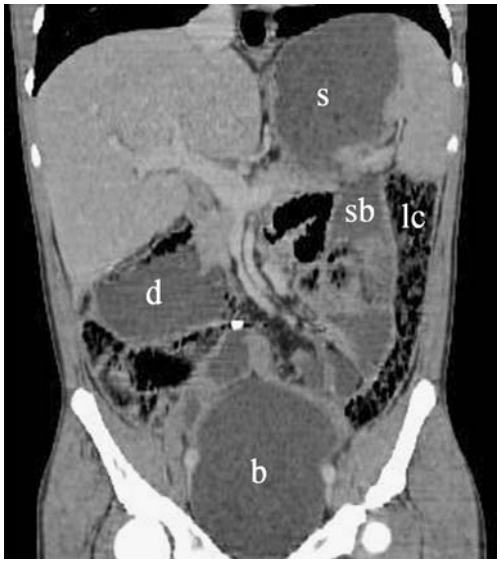

Fig. 4 A 19-year-old man with a known myogenic form of CIPO and prior history of ileostomy. The patient presented with abdominal pain and vomiting. Coronal CT reformation of MDCT shows marked dilatation of the stomach (s), duodenum (d), small bowel loops (sb) and left colon (lc) and excluded actual bowel obstruction. The patient was managed with conservative therapy. Distension of the bladder (megacystis) (b) is detected as a complication of the disease

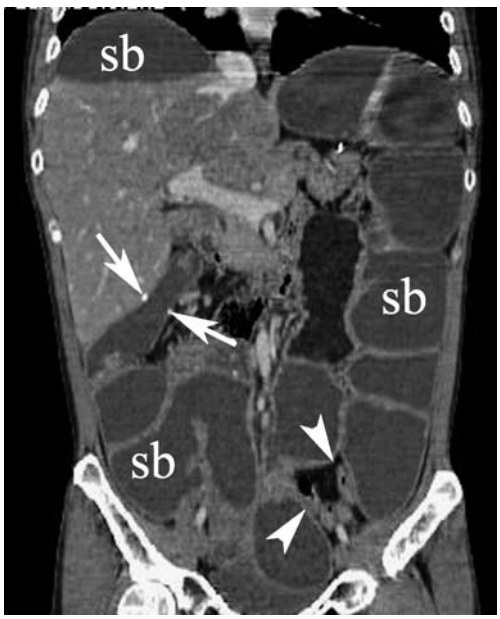

Fig. 5 A 49-year-old man with CIPO secondary to systemic disease who had prior subtotal colectomy and ileosigmoid anastomosis 3 years ago. The patient presented with abdominal pain, intestinal distension and vomiting, suggestive of stenosis at the anastomotic site. MDCT shows diffusely distended bowel loops (sb) and excludes actual mechanical obstruction at the level of the anastomotic site (arrows). No surgery was performed, and the patient was managed with conservative therapy

Fig. 6 A 51-year-old women with neurogenic form of CIPO of unknown origin with anti-Hu antibodies suggesting paraneoplastic syndrome. The patient presented with abdominal pain, nausea and vomiting. MDCT shows mild degrees of bowel distension and confirms the absence of actual obstruction. The distal small bowel loop is well seen, filled by feces (arrows), secondary to delayed transit time. The right portion of the colon is filled by feces. No underlying malignant tumor is seen. The symptoms disappeared with conservative therapy. No underlying neoplastic cause was found after a follow-up of 10 years

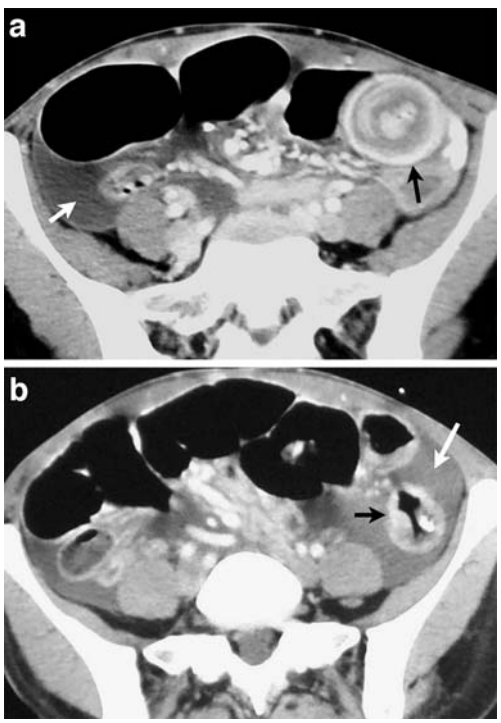

Fig. 7 A 40-year-old women with idiopathic CIPO. The patient presented acute abdominal pain requiring morphine therapy. **a** MDCT shows a "bowel-within-bowel" pattern, which is pathognomonic for enteroenteric intussusception (black arrow), but no lead-point tumor is identified, suggesting transient intussusception. Ascites is seen (white arrow). **b** Six months later, MDCT shows resolution of the intussusception in the corresponding area (black arrow). Ascites due to a low serum albumin level is still present (white arrow)

Because adult patients with CIPO have markedly decreased gastrointestinal motility, small bowel feces sign (Fig. 6), defined as particulate material and gas within dilated small bowel loops that simulate feces [24], may be seen. To our knowledge, there is no study in the literature that attempted to evaluate the frequency with which the small bowel feces sign can be depicted in adult patients with CIPO. However, in our experience, this feature is frequently observed and is associated with either mild or marked distension of the small bowel. In comparison with patients without CIPO, in whom the small bowel feces sign may indicate an actual small bowel obstruction [8], in patients with CIPO the small bowel feces sign does not correlate with actual small bowel occlusion.

Gastrointestinal complications

Intussusception Intussusception is defined by the prolapse of a bowel loop and its corresponding mesenteric fold into the lumen of an adjacent segment giving the so-called "bowel-within-bowel" classical pattern on MDCT imaging. Intussusception is incidentally observed on MDCT examinations performed in a non-selected population of patients [25], but as a rule, the absence of clinical symptoms of bowel obstruction, the absence of a demonstrable lead-point tumor on MDCT imaging and an involved segment of less than 3.5 cm in length are suggestive clues to the diagnosis of transient intussusception, which does not require surgery [25]. The frequency with which transient small bowel intussusception is observed in adult patients with CIPO is not known. Similarly, there is no study that allows knowing if intussusception is more common in patients with CIPO compared to patients without CIPO, since to our knowledge there is no report that describes this specific entity in the literature. However, patients with CIPO frequently complain of abdominal symptoms, so that when intussusception is seen on MDCT it may be difficult to make sure that it is actually a transient, self-limiting one, thus making the diagnosis more difficult to ascertain. In addition, because of the presence of bowel distension in patients with CIPO, it may be sometimes difficult to distinguish between actual bowel obstruction due to intussusception that would require surgery and transient self-limiting intussusception that resolves spontaneously. In such cases, a careful analysis of MDCT findings is mandatory to exclude the presence of a lead-point tumor

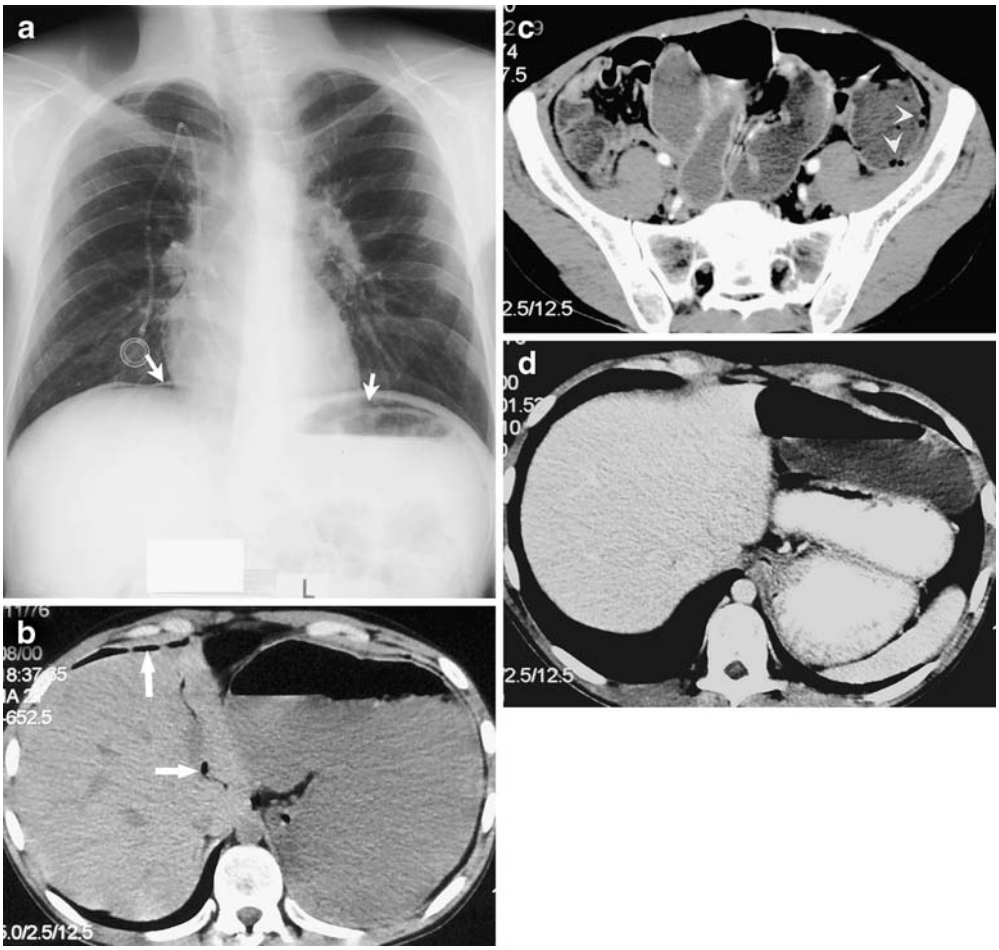

Fig. 8 A 23-year-old man with familial myogenic form of CIPO related to mitochondrial-neuro-gastro-intestinal encephalomyopathy. The patient presented with nausea, vomiting and abdominal pain irradiating to the left flank. **a** Chest film shows pneumoperitoneum (arrows). **b** Plain MDCT confirms the presence of a pneumoperitoneum (arrows) and shows gastric and duodenal distention. **c** At a lower level than in Fig. A, plain MDCT shows small air bubbles (arrowheads) in the left colon wall indicating pneumatosis. As neither signs of peritonitis nor inflammatory biologic changes were present, the patient was managed conservatively with rapid improvement of the clinical status. **d** One month later, MDCT confirmed complete resolution of the pneumoperitoneum

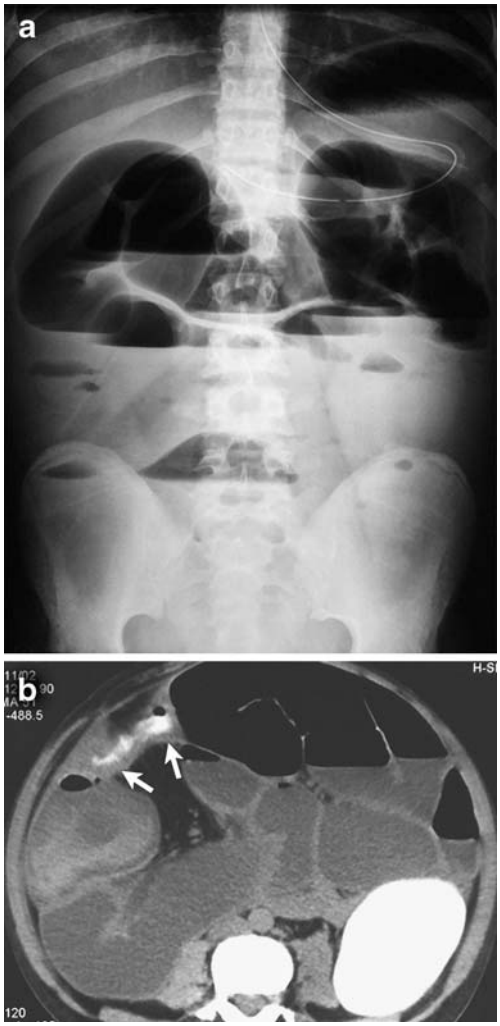

Fig. 9 A 20-year-old man with neonatal idiopathic CIPO and prior history of ileocolic anastomosis 14 years ago. The patient presented with nausea and vomiting. **a** Abdominal plain radiograph shows distended bowel loops and air-fluid levels, but does not help to rule out a mechanical cause for bowel obstruction. **b** MDCT shows a 5-cm-long stenosis (arrows) proximal to the anastomotic site. After resection of the stenotic portion, histopathologic findings were consistent with Crohn's disease

(Fig. 7). In difficult cases, and depending on the clinical status, a repeated and focused MDCT examination can be done the day after to confirm the transient and self-limited nature of the intussusception. This is critical to avoid unnecessary surgery in patients who have already had multiple operations.

Pneumatosis intestinalis Pneumatosis intestinalis is defined by the presence of gas within the bowel wall. Gas usually presents as small bubbles, but larger cysts may be seen. On abdominal plain radiography, follow-through studies and barium enema, it may be difficult to distinguish between gas within the gastrointestinal wall and intraluminal gas. In this regard, MDCT imaging is more discriminating than conventional imaging. Pneumatosis intestinalis is significantly more common in adult patients with CIPO compared to the normal population [26]. In such patients, pneumatosis intestinalis does not always indicate the need for surgical bowel resection and as a general rule does not correlate with bowel ischemia (Fig. 8) [26]. For this reason, in the presence of pneumatosis intestinalis, exploratory laparotomy should not be performed in the absence of peritoneal signs, fever and leukocytosis.

Superimposed diseases On a few occasions, MDCT helps to depict an associated gastrointestinal pathologic condition that may cause actual bowel occlusion (Fig. 9). In such cases, MDCT images help the physician to determine the most appropriate therapeutic option [7].

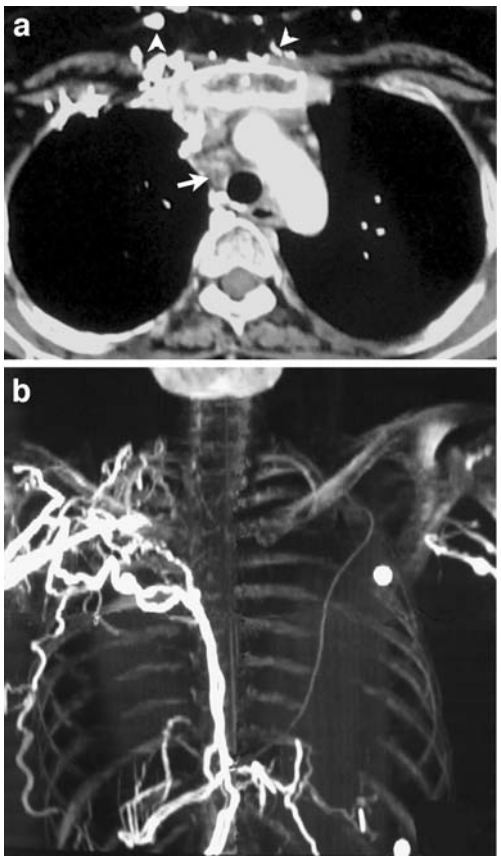

Fig. 10 A 33-year-old woman with myogenic form of CIPO related to mitochondrial cytopathy who had had subtotal colectomy and jejunocolic shunt 4 years ago. The patient was referred for intravenous nutritional therapy. **a** Axial MDCT of the thorax shows superior vena cava thrombosis (arrow) and numerous cavocaval pathways (arrowheads). **b** CT reformation using maximum intensity projection better shows the complex venous network

Extradigestive complications

Pneumoperitoneum MDCT is more sensitive than plain abdominal radiography to depict small amounts of extraintestinal gas. In a non-selected population of patients, it is well established that pneumoperitoneum indicates bowel perforation in 85% to 95% of the cases and requires prompt surgery [27, 28]. In the remaining 5% to 15% of cases, pneumoperitoneum is the result of a cause that does not warrant emergency surgery [28]. In adult patients with CIPO, pneumoperitoneum is more frequently a consequence of pneumatosis intestinalis and is observed in the absence of bowel perforation (Fig. 8) [25, 28]. Therefore, in patients with CIPO, MDCT imaging can show a pneumoperitoneum, that may be due to subserosal bubbles rupture into the peritoneum [26, 29]. For this reason, the presence of pneumoperitoneum in such patients must be integrated in a more comprehensive evaluation, including clinical examination of the abdomen to avoid useless surgery. In this situation, conservative management is required when no clinical features consistent with peritonitis are present.

Peritoneal fluid In adult patients with CIPO, peritoneal effusion, which can be incidentally depicted on MDCT imaging, may be secondary to a low serum albumin level due to malnutrition. The volume of free fluid may vary substantially among patients. Peritoneal effusion is mostly seen in patients who have an advanced stage of the disease (Fig. 7).

Other conditions Urinary tract involvement, which manifests as a bladder distension (also called megacystis) is the result of an impaired neuromuscular function (Fig. 4). In congenital CIPO, bladder involvement is found in 85% of patients with the myogenic form and 10% of patients with a neuropathic form [2]. The actual prevalence of bladder involvement in acquired CIPO is not known [2]. Long-term intravenous or parenteral nutritional therapy is frequently required to manage adult patients with CIPO as a consequence of multiple bowel resections. Complications of long-term intravenous nutritional therapy include hepatosteatosis, chronic diffuse hepatic disease, as well as deep or superficial venous thrombosis. Long-term intravenous nutritional therapy may lead to extensive thrombosis of the vein in which the intravenous device is placed [30]. MDCT imaging may be used to detect such complications and also to determine the extent of the disease (Fig. 10). This is helpful because deep venous thrombosis may be undetected by duplex Doppler ultrasonography. Also, venous thrombosis may be incidentally revealed by MDCT imaging. Because venous thrombosis may be responsible for fever in adult patients with CIPO, it is of importance to evaluate the patency of the vein in which an intravenous device is placed when MDCT imaging is performed to investigate fever of unknown origin.

Conclusion

MDCT shows a constellation of findings in adult patients with CIPO that are not obtainable with conventional imaging. MDCT helps suggest the disease in patients at their first presentation, and this is important since 80% of patients with CIPO are not correctly diagnosed at first presentation, and organic obstruction is the most frequent suspected diagnosis for which they have unappropriate surgery [11]. In such patients, MDCT provides pertinent and useful information regarding the caliber of the gastrointestinal tract and the status of the bowel wall, thus contributing to prompt and appropriate patient management. In patients with a known CIPO, the major role of MDCT is to rule out a mechanical cause of obstruction, and this role is perfectly fulfilled in the majority of cases, thus obviating the need for unnecessary laparotomy. In adult patients with CIPO, MDCT may show pneumatosis intestinalis, pneumoperitoneum or intussusception. However, these findings, which indicate the presence of several alarming pathological conditions that require urgent surgery in the general population, do not uniformly require surgery in patients with CIPO. Therefore, in such patients, the analysis of MDCT images must be integrated in a more comprehensive evaluation including the clinical status and the results of biological tests to avoid unnecessary surgery.

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