

## Case Report

# Chronic intestinal pseudo-obstruction in newborn: a case report

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### ABSTRACT

Chronic intestinal pseudo-obstruction (CIPO) is a rare and serious disorder of the gastrointestinal tract motility with the primary defect of impaired peristalsis. Symptoms are consistent with a bowel obstruction, although mechanical obstruction cannot be identified. It is a rare differential diagnosis for neonatal intestinal obstruction. Herein we report a case of neonate with non-resolving intestinal pseudo-obstruction, presenting since birth as progressive abdominal distention. The diagnosis was made by exclusion of mechanical causes of intestinal obstruction via thorough imaging studies.

**Keywords:** Impaired peristalsis, Intestinal obstruction, Motility disorder, Pseudo-obstruction

### INTRODUCTION

Chronic intestinal pseudo-obstruction (CIPO) is a rare and potentially lethal condition characterized by a severe impairment of gastrointestinal propulsion, leading to symptoms suggestive of partial or complete intestinal obstruction in the absence of any obstructive anatomical lesion. CIPO can involve any segment of the gastrointestinal tract; although the small bowel and colon are mainly affected. Clinical presentation is variable, and depends on the affected location.<sup>1</sup> CIPO is often considered as the most severe form of gut motility disorders; it comprises a heterogeneous group of conditions affecting the structure and/or function of the intestinal neuromusculature components.<sup>2</sup>

The etiology of pediatric CIPO is congenital in most cases, but the underlying mechanisms remain unclear. Because of etiologic heterogeneity, nonspecific manifestations, and low clinical awareness, the diagnosis of CIPO is likely to be delayed and the patients may suffer from multiple unnecessary surgical and medical measures, leading to high morbidity and mortality.<sup>3</sup>

### CASE REPORT

A preterm male infant was born at 35 weeks gestation via normal vaginal delivery to a healthy 24-year-old multipara mother. There was antenatal diagnosis of polyhydramnios and suspected fetal intestinal obstruction. There was history of consanguineous marriage. Apgar score was 8 and 9 at 1 and 5 minutes respectively. Birth weight was 2.4 kg, length was 45 cm and head circumference was 32 cm (All were at 50th percentile).

Upon admission to NICU the baby was stable with no dysmorphic features. There was marked abdominal distention associated with visible intestinal loops, visible peristalsis and thinning out of abdominal wall (Figure 1). Physical examination of other systems was unremarkable. The baby passed a small amount of meconium after rectal stimulation.

Plain x-ray of the abdomen revealed marked gaseous distension of the bowel loops down to the rectum, with no evidence of air-fluid levels (Figure 2).



**Figure 1: The baby showed marked abdominal distention with visible intestinal loops, and thinning out of abdominal wall.**



**Figure 2: Plain x-ray of abdomen showed marked gaseous distension of the bowel loops down to the rectum, and no evidence of air-fluid levels.**

Abdominal U/S showed normal liver, spleen, and both kidneys with no dilatation of the collecting systems. Gastrointestinal contrast studies (enema and meal) showed dilated and elongated bowel loops. The colon appeared elongated and tortuous with loss of colonic haustration. There was no evidence of intestinal obstruction (Figure 3 and 4).

Complete blood picture (CBC), blood chemistry, renal function tests and liver function tests all were normal. Blood C/S showed no growth. Stool for reducing substance was negative. Neonatal metabolic screening was unremarkable. Later, the baby started to pass regularly greenish watery meconium with no vomiting or

residual gastric aspirate. The abdominal distension became gradually less severe. On the 5th day of life orogastric tube feeding was started and advanced as tolerated.



**Figure 3: GIT contrast study (enema). The colon appeared elongated and tortuous with loss of colonic haustration.**



**Figure 4: GIT contrast study (meal) showed dilated and elongated bowel loops. There was no evidence of obstruction.**

Pediatric surgery consultation was done, and the case was thoroughly discussed; and a referral letter was sent to a higher center for further work-up and management. After one month of hospitalization, the baby was discharged home in good condition and after complete oral feeding was resumed. His weight was 2.2 kg at time of discharge. The baby was given follow-up weekly in our outpatient

clinic; waiting the response of the higher center. However, 12 days later the baby appeared in pediatric ED with a pronounced abdominal distension, marked irritability and respiratory embarrassment; but he was afebrile, passing stool and had no vomiting. On examination there were marked abdominal distension, dilated bowel loops with visible peristalsis and normal bowel sounds. He was hemodynamically stable. There was obvious failure to thrive and his weight was 1.7 kg. The baby was readmitted in NICU, isolation room. He was kept NPO and parental nutrition was commenced, full sepsis work-up was taken, and intravenous antibiotics were started. All cultures came back with no growth. Repeated radiological studies showed the same findings as in the first admission. Gradually, the condition stabilized and feeding was commenced after 3 days and increased gradually as the baby tolerated. Two weeks later the baby was transferred to the higher centre for further work-up and management. Their radiological studies were similar to ours. Fecal fat was absent. Sweat chloride test cannot be done (sweat cannot be collected); but cystic fibrosis transmembrane conductance regulator mutation analysis (CFTR mutation analysis) was done and the result was negative. The baby was managed by a multidisciplinary specialist team; who concluded that: the picture was compatible with the diagnosis of chronic intestinal pseudo-obstruction (CIPO). The baby was transferred back to our hospital with OGT special formula feeding, multivitamins and iron. His weight was 2.1 kg, and still had marked abdominal distension. Three days after readmission the baby developed two spikes of fever; so sepsis screening was taken and intravenous antibiotics were started. Fever was controlled and the baby became relatively stable. Again all cultures results came back negative. Eleven days later the baby developed another spike of fever, but he looked very toxic and dehydrated. The baby was resuscitated by nasogastric tube decompression, intravenous fluids and broad-spectrum intravenous antibiotics after taking full septic screening. As the hours passed, the condition was getting worse and sudden cardio-pulmonary arrest occurred; in spite of immediate active resuscitation, the baby was not revived and death was declared at age of about 82 days.

## DISCUSSION

Chronic intestinal pseudo-obstruction (CIPO) is a rare and debilitating condition. Patients usually present with severe impairment of gastrointestinal propulsion ensuing symptoms and signs suggestive of partial or complete intestinal obstruction in the absence of any mechanical lesion occluding the intestinal lumen. The vast majority of CIPO cases are sporadic and few cases are familial.<sup>4</sup> CIPO may be primary, when it exclusively involves intestinal smooth muscle or enteric neural plexus, or secondary to other conditions. The majority of pediatric CIPO cases are primary, with 65% to 80% of children becoming symptomatic by 12 months of age.<sup>5</sup> The CIPO histopathology can be divided into three forms;

neuropathic (the most common), myopathic and mesenchymopathic; based on abnormalities affecting the integrity of nerve pathways supplying the gut, smooth muscle cells or interstitial cells of Cajal, respectively.<sup>6</sup> Presentation may be acute or chronic and progressive, with symptoms suggesting partial or complete obstruction without the presence of obstructing lesions. Children may also present with urological involvement such as urinary retention and recurring urinary tract infections in addition to abdominal pain and distention.<sup>7</sup>

The diagnosis of CIPO is usually clinical and the work up is usually performed to rule out mechanical obstruction by means of radiologic studies. Our patient had serial plain abdominal radiographs and water soluble contrast studies that confirmed the patency of the intestine. Other investigations are usually performed to identify any underlying pathology leading to CIPO. Full thickness biopsies may provide diagnostic and prognostic answers.<sup>8</sup>

Prevalence and incidence of CIPO are widely unknown. A national survey in the USA reported that about 100 children are born with CIPO annually. A more recent survey in Japan found a prevalence of 3.7 per million children younger than 15 years with an equal gender incidence. These studies probably underestimate the actual number of new cases per year, because they do not include patients who develop CIPO symptoms later in life.<sup>9</sup>

There is no universally agreed protocol for management of CIPO in children, and there is wide variation in clinical practice. The management is directed to avoid unnecessary surgery, restore fluid and electrolyte balance, maintain an adequate caloric intake, promote coordinated intestinal motility, and treat complications such as sepsis and associated symptoms.<sup>10</sup> Treatment plans are complex and often necessitate the involvement of a large multidisciplinary team. Unfortunately, treatment options remain limited and success can be difficult to achieve. Therapy for secondary CIPO should focus on treating the underlying disorder.<sup>11</sup>

The prognosis is quite uncertain; most CIPO patients show a variable outcome, some cases may remain stable over time, whereas others rapidly worsen to unavoidable parenteral nutrition as a unique measure to prevent severe malnutrition and death.<sup>10</sup>

The nonspecific severe digestive symptoms, the suboptimal efficacy of medical treatments, and the pediatricians' limited knowledge of the disorder are some of the major factors contributing to the poor quality of life and the high rate of morbidity and mortality of CIPO patients.<sup>12</sup>

## CONCLUSION

Chronic intestinal pseudo-obstruction in neonates is rare and grave disorder. It must be considered in differential

diagnosis of neonatal intestinal obstruction. Radiographic studies were most helpful in making the diagnosis. Pediatric CIPO is still a challenging condition for clinicians, surgeons, and healthcare providers because of its complexity, heterogeneity, and different outcomes. The aim of this case report is to address the clinical importance and to raise pediatricians' awareness of this rare disorder.

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