

CLINICAL CASE

Congenital stenosis of the ileum in a 5-month-old infant with intestinal obstruction syndrome

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ABSTRACT

Background. Intestinal atresia refers to complete obstruction or partial stenosis of the intestinal lumen. Prevalence is 1/4000-5000 births, without gender predilection. When the blockage is incomplete, signs such as vomiting, abdominal distension, and constipation may occur shortly after birth or delayed in an unspecified manner. In intestinal atresia, the challenge may not be significant; however, stenosis may pose considerable difficulties. Obstructive syndrome must be ruled out in infant congenital intestinal stenosis. Even more rare cases have occurred during later ages.

Case Report. We report on a 5-month-old female infant with clinical symptoms of intestinal obstruction, which was managed surgically. Surgical finding was a congenital stricture of the ileum. The patient recovered without complications.

Conclusions. Obstructive syndrome in infant intestinal stenosis is a rare entity, which is usually not first suspected, but it should be ruled out as a study protocol for intestinal obstruction.

Key words: intestinal atresia, intestinal stenosis, intestinal obstruction.

INTRODUCTION

Intestinal obstruction syndrome is characterized by the interference with the flow of gases, liquids and solids and is clinically manifested by the triad of symptoms such as vomiting, abdominal distension and absence of bowel movements. Diagnosis is clinical and imaging techniques are used to locate the area of the obstruction. Obstructive lesions can be considered as intrinsic (e.g., atresia, stenosis, meconium ileus, etc.) or extrinsic (e.g., malrotation, strictures, etc.). They can also be classified according to the level of obstruction as high, medium or low (above the angle of Treitz, between the ligament of Treitz and the ileocecal valve or below it, respectively). The etiology of the obstruction is associated with the patient's age in most cases.¹⁻³

Intestinal atresia refers to a complete obstruction of the bowel lumen and stenosis refers to partial blockage of lumen. Its incidence is 1/4,000 to 5,000 births, without gender prevalence. Malformations of the small intestine constitute >90% of the cases. The most common cause of intestinal obstruction in newborns is intestinal atresia and in infants is intestinal invagination. Congenital ileal obstruction due to atresia or stenosis develops as a result of ischemia in any intestinal segment. Difficulty in diagnosis of intestinal obstruction due to intestinal stenosis increases when there is no clinical suspicion of the disease. Age at symptom onset is related to the degree of obstruction: the more constricted the intestinal lumen is, the earlier the symptoms will present themselves and vice versa. The differential diagnosis of vomiting in infants should include poor feeding techniques, pylorus hypertrophy and gastroesophageal reflux disease because these are the most frequent pathologies. Diagnosis is confirmed with the use of x-rays.⁴⁻⁶ Preferably, the lesion should be located before the intervention so as to guide the surgical approach. Macroscopic pathological features of all forms of intestinal atresia and stenosis are dilatation of the proximal segment to the obstruction and collapse of the small segment beyond the obstruction.⁵⁻⁸

This report presents the case of an infant with intestinal obstruction syndrome that was managed surgically, unexpectedly finding stenosis at the level of the ileum.

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The patient was a 5-month-old female. The patient's mother was a healthy 29-year-old with no gynecological/obstetrical complications. The patient was the product of the mother's first pregnancy. She was born vaginally, cried and breathed at birth without evidence of asphyxia, Apgar score 8-9, and with a birth weight of 3,200 g. She was discharged after 24 h without complications. She was breastfed and supplemented with formula from birth. She started weaning at 5 months. The mother stated that the patient had normal bowel movements since birth, with a frequency of one to three times every 24 to 72 h, with no history of constipation or other signs to indicate digestive pathology.

Four days prior, the patient experienced vomiting of gastrointestinal content and intolerance to oral feedings. During the previous 24 h she produced abundant nonbloody diarrheal stools with green mucous and with abdominal distension. Physical examination revealed an axillary temperature of 36.7°C, blood pressure 90/45 mmHg, weight 5,400 g (5th percentile), height 56 cm (<5th percentile), and generalized paleness. Cardiopulmonary auscultation was normal. Abdominal auscultation found a globular abdomen with peristalsis tension but unable to define any enlargements. The perineum and anus were intact. The remaining physical examination was normal. A nasogastric tube was placed and showed an abundance of gastric bile. X-rays showed dilated bowel loops and air-fluid levels in multiple segments (Figures 1 and 2).

We performed the following additional laboratory studies: urinalysis, coproparasitoscropy, fresh amoeba cultures, fecal mucus cytology, blood count and serum electrolytes. All results were normal; therefore, metabolic ileus and infectious process were ruled out and a surgical evaluation was requested. We decided to perform an exploratory laparotomy with a diagnosis of complicated intestinal obstruction.

During the procedure we observed significant strain loops and we found an area of 80% stenosis of the intestinal lumen at the level of the ileum at 50 cm from the ileocecal valve (Figure 3). We performed a resection of the stenotic area (8 cm) with full-enteroanastomosis end-to-end repair in two areas. Permeability was confirmed and the closing was performed smoothly. There were no histological alterations on the removed intestine and the patient evolved satisfactorily.

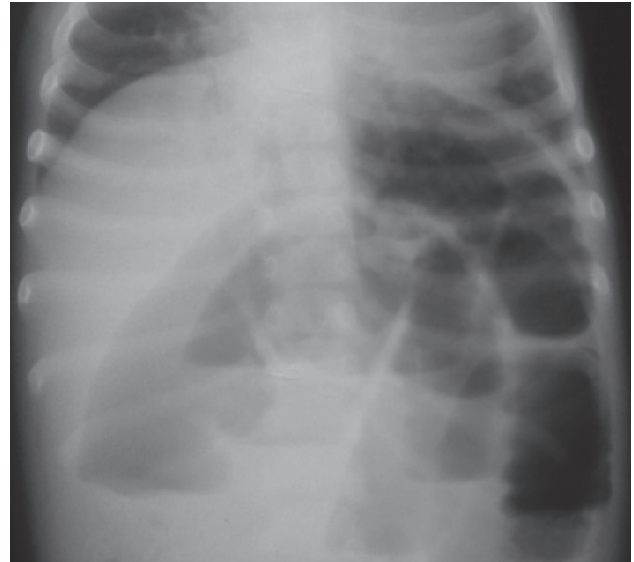


Figure 1. Anteroposterior x-ray of the abdomen on admission.

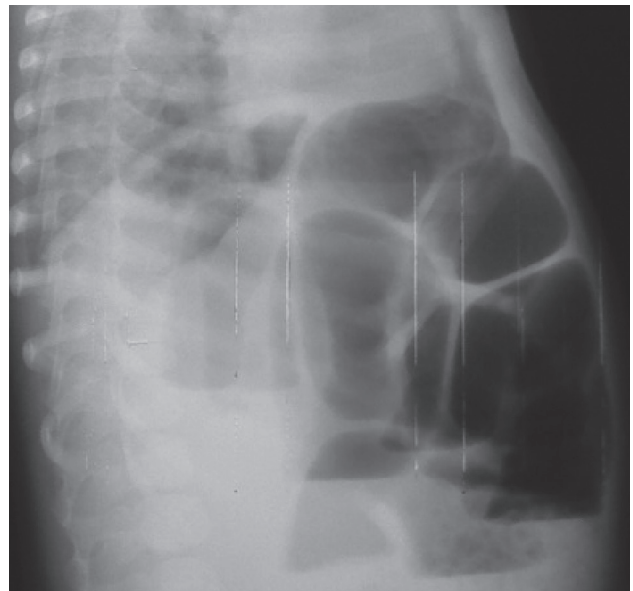


Figure 2. Right lateral abdominal x-ray on admission.

DISCUSSION

Congenital intestinal stenosis is a malformation of the bowel wall in which the proximal segment is dilated and is in continuity with the distal segment, existing in the junction between a short, rigid narrow segment that maintains a minimal lumen (Figure 4). The intestinal mesentery



Figure 3. Ileal stenosis at 50 cm from the ileocecal valve (surgical photo).



Figure 4. Dilated proximal stretch with rigid narrow segment that maintains a minimal lumen (surgical photo).

was intact and able to simulate its external appearance to that of a membranous atresia or type I, according to the classification by Grosfeld. Type I consists of the defect of the mucosa with the intact mesentery; type II represents a fibrous cord that connects the two atretic ends; type IIIa is a complete separation with a “V” defect in the mesen-

tery, type IIIb is an apple peel-like deformity and type IV represents multiple atresias (Figure 5).^{4,5,8}

Different hypotheses have been formulated regarding the origin of these atresias and of the intestinal stenosis. Initially, according to the Tandler theories, it was thought that they appear as a result of failures in the intestinal epithelial vacuolation. However, this idea was abandoned when it was shown that the intestinal lumen persisted throughout the entire embryonic development. Later, Louw (1959) and even later Louw and Barnard (1995) postulated that the main cause of the intestinal atresia is a vascular insufficiency of the bowel wall during fetal development, either a product of invagination, volvulus or incarceration.^{4,5,7}

Malformations of the small intestine occupy 95% of the cases of blockage in the intestinal tract during the neonatal period. Of these, the most common are the jejunum and ileum followed by duodenal atresia of 25 to 40% and multiple atresia occurring in 7 to 20% of patients.³⁻⁵ Different diagnostic imaging signs and laboratory tests such as the finding of polyhydramnios on a prenatal ultrasound, double-bubble image or alpha-fetoprotein levels may suggest intestinal atresia but are not as useful in the case of stenosis because the suspected diagnosis depends solely on clinical evaluation.⁶ Although the figures vary according to the type of literature, the authors generally

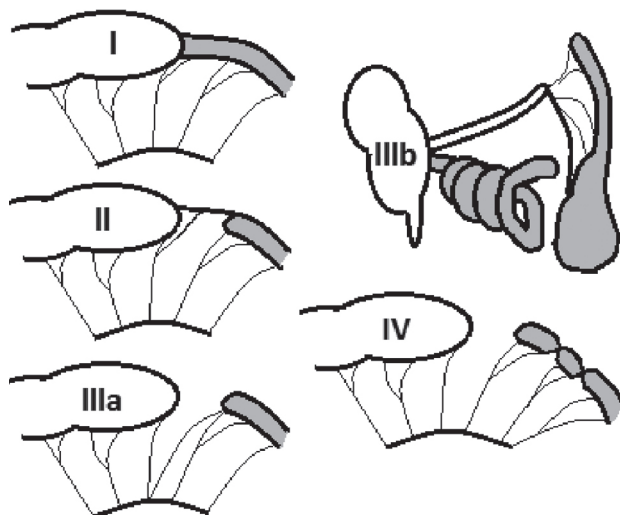


Figure 5. Grosfeld classification: I. Mucosal defect with intact mesentery. II. Fibrous cord that connects the two atretic ends. IIIa. Separation with defect in “V” of the mesentery. IIIb. Apple peel-like deformity. IV. Multiple atresias.

refer to the atresia and stenosis together, differentiated only by the degree of obstruction of the lumen and, hence, the clinical presentation.

In the infant, signs such as vomiting, abdominal distension and constipation may occur shortly after birth or delayed in an unspecified manner. Both atresia and severe stenosis have a clinical presentation of vomiting from the first days of life, abdominal distention to different degrees and, sometimes, an alteration in the passing of meconium. With stenosis, the manifestations are more passive and depend directly on the degree of obstruction; therefore, its diagnosis is delayed as happened in this patient who showed no gastrointestinal symptoms from birth until they appeared with a syndrome of intestinal occlusion. The only signs suggestive of this were low weight and height according to the percentile charts. We may also mention as a factor the onset of weaning, which had occurred 2 weeks prior to the illness. Based on a subtle clinical evaluation, the patient was not suspected of having any previous disease.

Congenital intrinsic atresia and stenosis are malformations with a common embryological origin. Although their differentiation is clearly pathological, clinical and radiological, such distinction can be very difficult.⁴⁻⁷ In this case, our patient presented with an intestinal obstruction syndrome that, by age group and according to her sudden clinical course offered no suspicion at first examination of intestinal stenosis (Table 1). As an unexpected finding and according to results of the laparotomy, it appeared that the treatment of choice for congenital intestinal stenosis was surgical resection of the stenotic segment, which could be performed via laparotomy^{8,9} with a favorable evolution. Postoperatively, the patient was examined and reported to be free of symptoms.

Table 1. Frequent causes of intestinal obstruction in lactating infants

- Invagination
- Hypertrophic stenosis of pylorus
- Incarcerated or strangulated inguinal hernia
- Meckel's diverticulum
- Volvulus
- Adhesions-postsurgical strictures
- Malrotation
- Paralytic ileus
- Intestinal stenosis-atresia

REFERENCES

1. Nerwich N, Shi E. Neonatal duodenal obstruction: a review of 30 consecutive cases. *Pediatr Surg Int* 1994;9:47-50.
2. Jiménez UP, Estrada MA, Gallego GJ. Estenosis duodenal congénita de presentación tardía: informe de tres casos. *Rev Mex Cir Pediatr* 2003;3:152-157.
3. Komuro H, Hori T, Amagai T, Hirai M, Yotsumoto K, Urita Y, et al. The etiologic role of intrauterine volvulus and intussusceptions in jejunoileal atresia. *J Pediatr Surg* 2004;39:1812-1814.
4. Stringer DA, Nadel H. Techniques for investigation of the pediatric gastrointestinal tract. In: Stringer DA, Babyn PS, eds. *Pediatric Gastrointestinal Imaging and Intervention*. Hamilton, Ontario, BC: Decker; 2000. pp. 15-74.
5. Langer JC. Abdominal wall defects. *World J Surg* 2003;27:117-124.
6. Swischuk LE. *Radiología en el niño y en el recién nacido*. Madrid: Marbán; 2005. pp. 541-544.
7. Shalkow J, Quiros JA, Shorter N. Small intestinal atresia and stenosis. *eMedicine Journal* (online) 2004. Available at: <http://emedicine.medscape.com/article/939258-overview>
8. Dalla Vecchia LK, Grosfeld JL, West KW, Rescorla FJ, Scherer LR, Engum SA. Intestinal atresia and stenosis: a 25-year experience with 277 cases. *Arch Surg* 1998;133:490-497.
9. Strauch ED, Hill JL. Intestinal atresia. In: Ziegler M, Azizkhan G, eds. *Operative Pediatric Surgery*. New York: McGraw Hill; 2003. pp. 590-593.