Colonic atresia: a rare entity in the newborn. A six-case report and a bibliographic review

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ABSTRACT

Introduction. Colonic atresia is a rare malformation accounting for 1.8-15% of all intestinal atresias. We present a 6-case series along with a bibliographic review.

Clinical case. This 6-case series consists of three female cases and three male cases diagnosed 24-84 hours following birth. They were all located in the right hemicolon. The most relevant clinical signs included abdominal distension, absence of defecation, and bilious to fecal vomit. Repairs included primary or step-by-step anastomoses for bowel transit reconstruction. One patient diagnosed at > 72 hours of life died.

Conclusion. In our experience with colonic atresia, when primary or step-by-step intestinal recanalization (diversion for future repair) is achieved, the expected prognosis is excellent, provided that colonic atresia has been diagnosed at 24-48 hours of life.

KEY WORDS: Colonic atresia; Intestinal obstruction; Newborn; Anastomosis; Intestinal diversion.

ATRESIA DE COLON, UNA ENTIDAD POCO FRECUENTE EN EL RECIÉN NACIDO. A PROPÓSITO DE SEIS CASOSY REVISIÓN BIBLIOGRÁFICA

RESUMEN

Introducción. Las atresias de colon son malformaciones raras que comprenden alrededor de 1,8-15% de total de las atresias intestinales. Presentamos una serie de seis casos junto con una revisión bibliográfica.

Caso clínico. Esta serie de casos incluye tres casos femeninos y tres casos masculinos que fueron diagnosticados entre 24-84 horas del nacimiento. Todas se localizaron en el hemicolon derecho. Las manifestaciones clínicas más relevantes fueron distensión abdominal, ausencia de evacuaciones, vómitos de biliosos a fecaloideos. Las correcciones incluyen anastomosis primarias o por etapas para la reconstrucción del tránsito intestinal. Se presentó una defunción en un paciente diagnosticado con > 72 horas de vida.

Conclusión. En nuestra experiencia, en la atresia de colon, cuando se logra recanalizar el intestino de forma primaria o por etapas (derivación para la futura corrección), se espera excelente pronóstico, siempre que sea diagnosticada entre las 24 a 48 horas de vida.

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Date of submission: June 2020 *Date of acceptance:* October 2020

PALABRAS CLAVE: Atresia de colon; Obstrucción intestinal; Recién nacido; Anastomosis; Derivación intestinal.

INTRODUCTION

Colonic atresia (CA) is a cause of gastrointestinal tract obstruction in the newborn (NB). It is defined as a congenital loss of continuity in a colonic segment[1,2]. It is a rare malformation, with an incidence of 1 out of 20,000 live newborns (LN), and it accounts for 1.8-15% of all intestinal atresias. It represents a neonatal surgical challenge for the pediatric surgeon[1-6].

Given how rare CA is, no large series have been reported in the literature yet. CA can occur in an isolated fashion or associated with abnormalities in other systems[7].

The first case was reported by Bininger (1673). In 1922, Gaub published the first surviving child, who underwent proximal diversion colostomy. The first report on definitive treatment (anastomosis) in children was described by Potts (1947)[4,8,9].

We present 6 cases of colonic atresia, with different surgical repair techniques. The pathology is so rare the authors would like to spread the word about it.

CASE REPORTS

This series consists of 6 CA cases managed at Dr. Robert Reíd Cabral Pediatric Hospital in Santo Domingo (Dominican Republic) from 2014 to 2018. They were catalogued as intestinal obstruction at 24-84 hours of life. Three patients were female and three patients were male. All atresias were located in the right hemicolon. There were four type III CAs, one type II CA, and one type I CA. Simple abdominal X-ray showed intestinal obstruction signs, with five patients presenting a large dilated loop in the right hemiabdomen.

Table 1 features patient characteristics, imaging findings, surgical procedures, and re-interventions. Figure 1

Table 1. Colonic atresia cases managed at Dr. Robert Reíd Cabral Pediatric Hospital (Dominican Republic) from 2014 to 2018.

No.	*Sex*	*Age*	*Weight*	*Clinical signs*	*Type of atresia*	*Radiological findings*	*Surgery*	*Re-intervention*	*Complications*
1	Fem	2 days	2.9 kg	Bilious vomit. No defecation. Abdominal distension	Dilated proximal colon, segment until hepatic flexure separated from the distal segment of the microcolon from the transverse colon. TYPE III	Hydroaerial levels at the level of the thin loops. No distal air	Devine's colostomy	At 1 year of age. Resection and end-to-end colonic anastomosis	None
2	Fem	1 day	2.9 kg	Feeding refusal. No defecation. Abdominal distension	Dilated proximal colon. Right third segment of the transverse colon separated from the distal segment of the descending colon. TYPE III	Evidence of highly dilated gastric pouch with another superimposed image of right bowel dilatation. No distal air	Enteroplasty + side-to-side colocolonic anastomosis	No	None
3	Male	2 days	3.4 kg	Bilious vomit. Abdominal distension. No defecation	Dilated proximal colon, ascending segment until hepatic flexure separated from the distal segment of the transverse colon. TYPE III	Hydroaerial levels Image of great loop dilatation on the right side	Enteroplasty + end-to-side ileocolonic anastomosis	Ileostomy + colostomy distal segment. At 2 years of age: ileocolonic anastomosis	Anastomotic dehiscence with widespread peritonitis (4 days of age)
4	Fem	3 days	3.2 kg	Hypoactive. Abdominal distension. No defecation	Dilated proximal ascending colon before hepatic flexure. TYPE III	Hydroaerial levels with large image of dilated loop on the right side	Devine's colostomy	At 2 years of age. Resection and end-to-end ileocolonic anastomosis	Adhesion syndrome two weeks following recanalization
5	Male	4 days	3 kg	Fecal vomit. No defecation. Abdominal distension. Abdominal sensitivity. Moderate to severe dehydration	Perforated proximal ascending colon, continuity with a hypoplastic segment with intraluminal membrane and full mesentery. TYPE I	Intestinal loops displaced towards the left hemiabdomen with 2 air bubbles in the right hemiabdomen and great radio-opacity. Free fluid at the abdominal cavity without distal air	Ileostomy with mucocutaneous fistula	No	Died as a result of a septic shock on postoperative day 2
6	Male	2 days	3.6 kg	No defecation. Abdominal distension. Feeding refusal. Bilious vomit	Proximal ascending colon ending in a blind pouch, distal end 2 cm away joined by the mesocolon. TYPE II	Dilated intestinal loops, loop with the greatest diameter on the right side. Standing X-ray shows hydroaerial level of the right hemiabdomen, without distal air	Dilated proximal colon resection. End-to-end ileocolonic anastomosis	No	None

Source: Pediatric Surgery Department, Dr. Robert Reíd Cabral Pediatric Hospital, Santo Domingo (Dominican Republic)

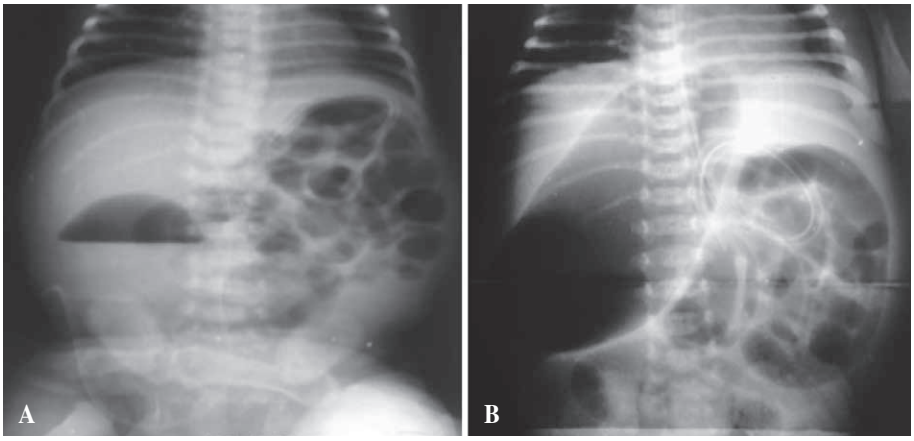

Figure 1. A) Simple standing X-ray. The arrow indicates a single hydroaerial level in the right hemiabdomen. B) A greatly dilated loop can be seen in the right hemiabdomen; it could be mistaken for a pneumoperitoneum. The arrows indicate radiolucency's extension.

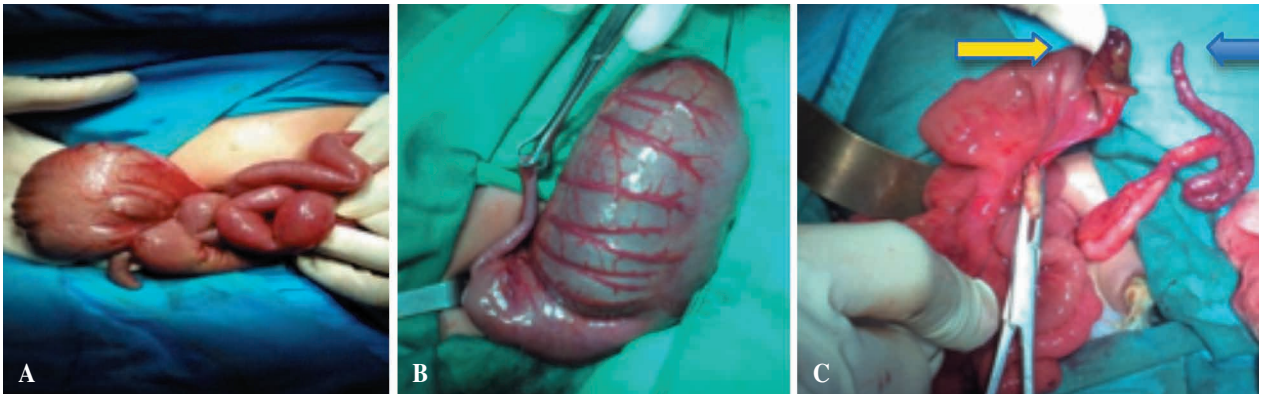

Figure 2. Type III right colon atresias: A) Proximal ascending colonic atresia, with significant ileal dilatation; B) Proximal ascending colonic atresia with vermiform appendix; C) Proximal colonic atresia (yellow arrow) and hyperplastic colonic segment (blue arrow).

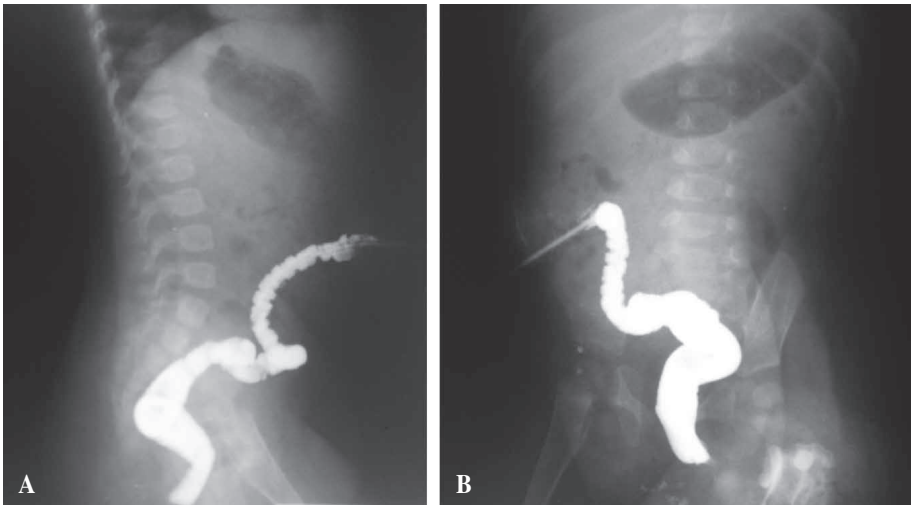

Figure 3. Distal colostogram: A) Lateral projection; B) Oblique projection, in a patient with type III colonic atresia, at 2 years of age, following intestinal reconstruction where increased distal colon diameter can be seen, which allows for anastomosis.

shows simple X-ray images, Figure 2 demonstrates the morphology of certain atresias, and Figure 3 displays the contrast-enhanced colon following intestinal recanalization.

Generally speaking, clinical signs included abdominal distension and absence of defecation. Four patients had bilious vomit, one patient had fecal vomit, and there were also some instances of feeding refusal, hypoactivity, dehydration, etc.

Colostomy was performed in two NBs while exteriorizing both sides for subsequent repair. One NB with

cutaneous fistula of the distal colon underwent ileostomy, and 3 NBs underwent primary anastomosis – one of them required re-intervention and step-by-step repair 4 days following surgery as a result of anastomotic dehiscence with widespread peritonitis.

One death was recorded in our series – a patient with type I CA referred to our unit after the first 72 hours of life who received assisted mechanical ventilation and died as a result of a septic shock. The three deferred bowel transit reconstructions were performed at 1-2 years of age. One of these patients had adhesion syndrome and underwent adhesiolysis. Patients had a favorable evolution following definitive surgical repair, with an adequate defecating pattern.

DISCUSSION

Colonic atresia is considered the most infrequent form of gastrointestinal tract atresia. Various hypotheses on the origin of intestinal atresia have been proposed: Tandler[2] pointed to the lack of vacuolization of the intestinal epithelium in intestinal development. Louw and Bernard (1956) argued it is due to the presence of an ischemic lesion (fetal mesenteric vessels), once the mid bowel has returned to the coelomic cavity, as a result of intrauterine invagination and volvuluses after the embryonic period. Maternal vasoactive drug intake has also been proposed to potentially increase the risk of developing this malformation[1,9,11,12]. Today, there is new evidence suggesting that genetics may also play a role in CA pathogenesis[3,10].

In a study, Etensel et al. reported that 3.7% of intestinal atresias were CAs, while Dalla Vecchia et al. found 277 intestinal atresia cases in 25 years, 15 of which were CAs[2]. Davenport described 118 infants with colonic atresia, with 28% of the lesions occurring in the ascending colon, 3% at the hepatic flexure, 23% in the transverse colon, 25% at the splenic flexure, and 20% in the descending and sigmoid colon[4,7]. In our series, all CAs occurred in the right hemicolon – 50% in the ascending colon, 33% at the hepatic flexure, and 17% in the right proximal third of the transverse colon. Considering our healthcare facility is a third-level, national reference hospital in the Dominican Republic, anatomical location did not demonstrate any relationship or factor associated with the pathology itself.

CA is associated with fewer abnormalities than other atresias, but they still can impact prognosis. Abnormalities include exophthalmos, bilateral optic nerve hypoplasia, bladder exstrophy, omphalocele, gastroschisis (2.5-3%)[5,6], intestinal malrotation, and intestinal atresia in up to 20% of cases[13]. A combination of imperforate anus and CA has been reported in six patients in the British literature[11,14]. 2% of CA patients have Hirschsprüng's disease (HD)[4,15]. Congenital heart abnormalities, polydactyly, syndactyly, and lack of radius have also been described[4-6]. Our series features 6 cases of isolated colonic atresia.

The most widely used classification divides colonic atresia into three types: type I is mucosal atresia with intact intestinal wall and mesentery. In type II, the atretic ends are separated by a fibrous cord. In type III, the atretic ends are separated by a V-shaped mesenteric space. Type III lesions are the most frequent, whereas types I and II are more distal to the splenic flexure[2,4,6]. All types of atresia cause potentially great proximal segment dilatation, while the bowel, beyond discontinuity, has a small diameter (hypoplastic[4-6]. However, this classification system has limited clinical value because surgical principles and results are not based on the anatomical categorization of the lesion[4]. 67% of our cases were type III.

CA clinical characteristics include abdominal distension – which can be marked and progressive at 24-48 hours of life – late vomit – on the second or third days of life –, and inability to eliminate the meconium present in distal bowel obstruction[2-6,13]. Even though CA can be diagnosed prenatally using ultrasonography, diagnosis is usually suspected at birth[6,11,12]. In this case series, there were no prenatal diagnoses.

Simple abdominal X-ray in CA demonstrates air-fluid levels. Although the small bowel and the colon cannot be easily identified in the neonatal period, the distension proximal to the obstruction is typically greater in the colon. It looks like "emerald crystal" and it is so massive it may have the appearance of a pneumoperitoneum – the presence of pneumoperitoneum is a sign of proximal colon perforation[4,6,9,11,16]. Diagnosis is achieved using a contrast enema, which demonstrates a distal colon with a small diameter interrupted at the level of the obstruction. In addition, other causes of distal obstruction such as malrotation, HD, meconium ileus, etc. should be identified. Once CA has been diagnosed, surgical management should be urgent, since the risk of perforation is higher[4-6,13,17,18]. In 5 of our cases, simple X-ray showed a large dilated loop corresponding with the colon in the right hemiabdomen.

Surgical treatment involves either resection and primary end-to-end anastomosis, or a step-by-step approach, with intestinal diversion such as terminal colostomy, and in the presence of mucosal fistula with subsequent recanalization, other techniques such as Swenson and Santulli procedures, which subsequently require stoma closure[2,3]. Suction rectal biopsy is recommended at primary or step-by-step surgery for HD diagnosis, which is usually achieved following anastomotic failure when restoring intestinal continuity[4-6,15,19].

Our patients underwent various techniques such as end-to-end anastomosis, enteroplasty with side-to-side anastomosis, end-to-side anastomosis with subsequent dehiscence in one case, and diversion of both segments – the proximal segment for removal purposes, and the distal segment for water injection purposes in order to increase the diameter

and the intestinal lumen of the hypoplastic segment – in three cases. Intestinal continuity was achieved at 1-2 years of age as a result of inadequate patient follow-up, with patients presenting again at surgery consultations after 1 year of age.

It should be mentioned that rectal and anal atresia are not included in CA – they belong to the anorectal malformation spectrum, representing 1%[20].

Mortality is < 10% if adequately managed and in the absence of other pathologies. However, late diagnosis beyond the first 72 hours of life can potentially be associated with a > 60% mortality[3,4]. In our series, one patient with type I atresia diagnosed at 84 hours of life died.

CONCLUSION

Colonic atresia has an excellent prognosis provided that bowel recanalization is achieved. Caliber differences between the proximal (dilated) and the distal (hypoplastic) ends to the atresia should be highlighted. The challenge is to achieve primary bowel continuity. When proximal and distal bowel diversion is required as a result of caliber differences at both ends, the distal colon can be irrigated with water or saline solution to increase diameter and intestinal lumen for subsequent bowel transit repair and restoration. With an early diagnosis and an adequate management, results are satisfactory.

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