Journal of Neonatal Surgery 2018; 7:22
DOI: 10.21699/jns.v7i2.743

OPEN ACCESS

ORIGINAL ARTICLE

Atresia of the Colon: Etiological Aberrations, Clinical Observations, and Challenges in Management

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How to cite: Pant N, Singh S, Rawat J, Kureel SN, Wakhlu A. Atresia of the colon: Etiological aberrations, clinical observations, and challenges in management. J Neonatal Surg. 2018;7:22.

ABSTRACT

Objective: The objective of this study is to review the clinicoradiological profile, scheme of management and the outcome in cases of colonic atresia (CA), and ascertain an optimal approach for the treatment of CA to minimize morbidity and mortality. **Design and Setting:** This was a retrospective observational study carried out at a tertiary health-care center. **Duration:** Total of 6 years duration (January 2011–December 2016). **Materials and Methods:** A retrospective analysis of 10 patients of CA managed over a 6- year period. Data related to demographics, clinical presentation, associated anomalies, radiologic, intraoperative findings, postoperative stay, complications, and outcome were analyzed. **Results:** There were three cases of Type II atresia involving terminal ileum, cecum, and adjacent colon. Three cases had proximal ascending colon atresia (Type IIIa [*n* = 2]; Type I [*n* = 1]), and two cases of type IIIa atresia of the hepatic flexure. Two babies had atresia involving the sigmoid colon; one had Type II atresia, while we were unable to assign a type to the other within the prevailing classification. Seven babies were initially treated with a stoma either in the ileum (*n* = 3), hepatic flexure (*n* = 2), and sigmoid colon (*n* = 2), whereas three were treated with a primary anastomoses. Cases treated with a primary anastomoses had lesser morbidity and a better outcome than those with an ileal or ascending colon stoma. **Conclusion:** Contrary to the theory of an acute antenatal vascular accident, CA may rarely result from a gradual, sequential obliteration of mesenteric vasculature. Primary anastomosis should be contemplated in proximal CA wherever possible as stomal complications, especially high stoma output can result in considerable morbidity.

Key words: Colonic atresia; Dehydration; Gastrografin enema; Intestinal stoma; Mesenteric vascular occlusion

INTRODUCTION

Colonic atresia (CA) is a less common site for intestinal atresia. We report 10 cases of CA managed over a 6-year period and describe the presentation, diagnosis, management, and outcome of these patients. An attempt has been made to speculate on the likely embryogenesis of an unusual variant of CA. We aim to put forward the management options for patients with CA with a view to suggest a protocol to minimize morbidity and mortality.

MATERIALS AND METHODS

The material for this study comprised 10 cases of CA admitted and managed between 2011 and 2016. The patients' records were studied and data related to demographics, clinical presentation, associated anomalies, radiologic findings, intra-operative findings, duration of postoperative stay, complications, and outcome of the patients was analyzed. At our institution, the protocol is to routinely do a gastrografin enema in suspected cases of uncomplicated, low intestinal obstruction in neonates. Very sick babies with underlying peritonitis/perforation were treated initially with a bilateral peritoneal drain (PD) inserted under local anesthesia (LA) until their condition improved. At surgery, determination of the type of atresia (as per Grosfeld classification) as well as the site and extent of involvement of colon was done [1]. Wherever possible, a primary anastomosis was performed, but the decision to perform an anastomosis or a diverting stoma was essentially an intraoperative decision based on the condition of child, site of the CA, and the status of the bowel. The dilated segment

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Submitted: 12-02-2018 Accepted: 20-03-2018
Conflict of interest: None Source of Support: Nil

proximal to the atresia was excised while doing either a primary anastomoses or a stoma. Rectal biopsy to rule out Hirschsprung's disease (HD) was not performed in any child. In cases where a stoma was constructed at primary surgery, a distal colostogram was performed to check distal patency before stoma closure. Our protocol for patients with a stoma is closure after 6–8 weeks of primary surgery, subject to the condition that these babies have an adequate weight gain and a proper nutritional and hydration status. We routinely prescribe (ORS-zinc solution) oral rehydration solution supplemented with zinc to all neonates with a stoma till the time of stoma closure to compensate for their high stoma losses and minimize chances of dehydration. ORS has been used as a supplement to maintain water and electrolyte balance in infants with ileostomy and in patients with high ileostomy fluid output [2,3]. Follow-up of patients was recorded up to the last outpatient visit. Follow-up interval was between 6 and 15 months.

When the atretic part included the terminal ileum, cecum, and adjacent ascending colon, we used the term ileocolic atresia to differentiate them from cases where the atresia involved part of the ascending colon with a small proximal ascending colon stump, which were termed as atresia of proximal ascending colon.

RESULTS

Over the period of study, 85 cases of intestinal atresia were managed out of which 10 (11.8%) had atresia involving some part of the colon. There were 7 male and 3 female babies. Four were in-hospital births, whereas 6 cases were referred from other hospitals. The median birth weight was 2.5 kg (range 2–3 kg). Eight patients had third-trimester antenatal ultrasound (US) scan out of which 4 had both second- and third-trimester scans. A dilated echogenic bowel was mentioned in only one report. Apart from one patient who had an associated anorectal malformation (ARM), no other patient had an associated anomaly or a syndromic association. Seven cases were operated within the first 3 days of life, whereas three were operated on the 5th, 8th, and 12th day, respectively, in view of late presentation/low general condition.

Eight out of the 10 (80%) cases of CA involved the right half of the colon, while only 2 involved the left side. Of the 8 right-sided CA, the sites included- 3 cases of ileocolic region (all type II), 3 cases of proximal ascending colon (2 type IIIa and 1 type I), and 2 cases of hepatic flexure (both type IIIa). Two babies had atresia involving the sigmoid colon; one had Type II atresia, while we were unable to assign a type to the other within the prevailing classification.

Four cases were operated without a preoperative gastrografin enema (Table 1). These included cases 1, 2, and 3 who had features of intestinal perforation and peritonitis and had presented late in poor general condition. They were initially managed with PD (cut out from a sterile surgical glove) inserted under LA in the ward. Case 4 was initially diagnosed as a case of high ARM based on an absent anal opening and a relatively flat perineum. The distal gas shadow on X-ray was very high in the pelvis. At surgery, the entire colon was found devoid of its mesentery up to the mid-sigmoid region with the absence of the ileocolic, right colic, middle colic arteries and the sigmoid branches of the left colic artery. Instead, the colon was supplied by a single marginal artery which was a continuation of a terminal ileal artery (Figure 1). There were no identifiable tinea coli and haustrations, and the entire proximal colon was filled with meconium except for the last 1 cm, which was narrow. There was no pouch-like dilatation of colon on X-ray. Second, no colonic pouch found intraoperatively excluding the possibility of congenital pouch colon. As earlier mentioned, the perineum was flat and no anal opening was present that exclude the diagnosis of rectal atresia.

The distal rectosigmoid colon was akin to a microcolon up to the peritoneal reflection. The remaining six patients presented with clinical features and radiologic features suggesting a distal bowel obstruction and a contrast enema was performed in these six babies. Features on contrast enema diagnostic of CA in some of these cases are shown in Figure 2 and Figure 3a and b and the operative findings are shown in Table 2. Seven babies were managed initially by a diverting stoma while a primary end-to-oblique anastomosis was performed in three patients. Average postoperative hospital stay was 10 days (range 7–19 days). Four babies with a stoma developed stomal diarrhea and needed readmission. There were 2 deaths (mortality 20%). The patient with CA and associated ARM died on the 8th postoperative day due to sepsis. Another baby admitted twice for stomal diarrhea died 3 days after the 2nd admission due to dehydration, dyselectrolytemia, and malnutrition. The remaining five babies with a stoma have had their stoma closed between 3 and 4 months of primary diverting surgery. This delay was due to a slow weight gain secondary to dehydration, dyselectrolytemia, and poor nutrition status in the initial months after surgery. Following stoma closure, these patients fared well. Furthermore, the three patients who had a primary anastomosis had a good outcome till their last outpatient visit.

DISCUSSION

CA is a rare condition comprising 1.8–15% cases of intestinal atresia and constituted 11.8% of all intestinal atresia in our series [4]. The condition is classified into two broad clinical groups, isolated CA (found in about 2/3 of cases) and CA associated with abdomi-

Table 1: Presentation and management of the 4 cases of CA operated without a contrast enema

Case	Presentation	X-Ray	Preoperative treatment	Laparotomy Findings	Surgical Procedure
1	Late presentation. Distended, shiny, erythematous abdomen. Shock	Not done	Intravenous fluids. Vasopressors. Antibiotics. PD under LA.	Type II atresia of sigmoid colon with perforation proximal colon	Divided sigmoid colostomy
2	Abdominal distension, constipation.	Pneumoperitoneum. Multiple dilated bowel loops.	Intravenous fluids. Antibiotics. PD under LA.	Type I proximal CA with gangrene and perforation terminal ileum.	Excision of ascending colon stump, cecum, and dilated terminal ileum. Divided ileocolostomy
3	Abdominal distension, constipation. Shock	Pneumoperitoneum.	Intravenous fluids. Antibiotics. PD under LA.	Type II ileocolic atresia. Distended terminal ileum with perforation.	Excision of the distended terminal ileum. Divided ileocolostomy
4	Abdominal distension, absent anal opening.	Prone cross table X-Ray suggestive of high ARM	Intravenous fluids. Antibiotics.	An associated sigmoid CA with the absence of major colonic vessels.	Divided colostomy.

CA: Colonic atresia, PD: Peritoneal drainage, LA: Local anesthesia, ARM: Anorectal malformation

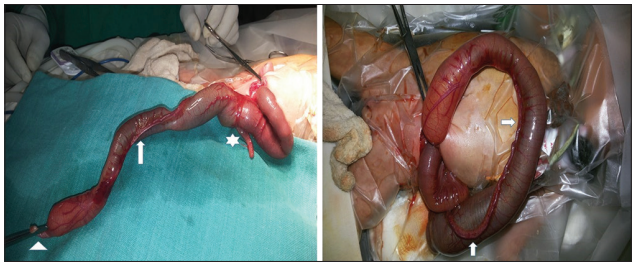

Figure 1: Entire colon, devoid of its mesentery, tinea coli, is supplied by a single marginal artery (white arrow). The terminal portion of distal most segment of proximal colon is hypoplastic (white arrowhead). Well-formed cecum and appendix (white star)

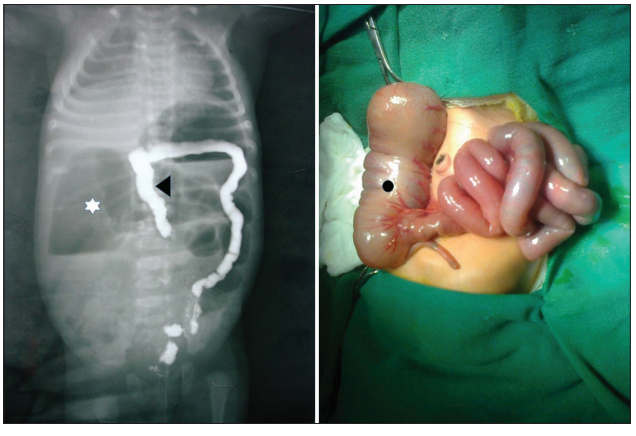

Figure 2: Contrast enema film showing dilated ascending colon with an air-fluid level (white star). It occupies most of the right half of abdomen and turns the right half of unused transverse colon on itself (black arrowhead). Intra-operative photograph of the same patient, revealing a type IIIa atresia at hepatic flexure. Both cecum and ascending colon are not retroperitonized (Black dot)

nal wall defects, other intestinal atresia, and/or malrotation. Rare associations include ARM, esophageal atresia, or HD [4]. Isolated CA is reportedly seen in full-term, good birthweight babies and has an excellent outcome, whereas the outcome is not so good in CA associated with other anomalies [4-7]. The babies in our series had a median weight of 2.5 kg and were full- term. In nearly 80% of our cases, the site of atresia was proximal to the mid-transverse colon which is similar to the figure of 83.3–86.6% incidence of proximal CA reported in literature [4-6]. A literature survey reports that nearly 47.3% cases of CA have one or more congenital anomaly, namely, gastrointestinal, abdominal wall defect, and malrotation [4]. This means nearly 52.7% cases are isolated atresia [4]. However, in our series, 90% patients had isolated CA. The experience of different series repoorted from Indian subcontinent has been varied. A series from India reported 50% associated anomalies in 4 cases of CA, whereas another series reported no association in the 4 babies of CA. [7,8]

The genesis of intestinal atresia is generally attributed to an acute antenatal vascular accident, secondary to an extrinsic bowel obstruction with resultant aseptic necrosis and resorption [9]. Considering that not all mesenteric vessel obliterations could be attributed to extrinsic obstruction, cases of familial atresia were ascribed to specific embryologic pathways that, when disrupted, give rise to similar anomalies [10]. Other

Table 2: Presentation and management of the six cases operated following a contrast enema

Case	Surgical findings			Primary procedure performed
	Site of atresia	Type of atresia	Condition of bowel	
1	Ileocolic	II	Grossly dilated ileal atretic segment Proximal ileum was congested and red Unsuitable for a primary anastomoses	Excision of dilated ileal segment Divided ileocolostomy
2	Proximal ascending colon	IIIa	Condition of the proximal and distal bowel healthy	Excision of ascending colon stump, dilated cecum, proximal dilated ileum, and 1 cm of the distal atretic colon End-to-oblique ileocolic anastomosis
3	Hepatic flexure	IIIa	Grossly dilated ascending colon up to hepatic flexure Terminal ileum of normal caliber	Divided stoma of ascending and transverse colon
4	Proximal ascending colon	IIIa	Condition of bowel healthy	Excision of ascending colon stump, dilated cecum, proximal dilated ileum, and 1 cm of distal atretic colon End-to oblique ileocolic anastomosis
5	Hepatic flexure	IIIa	Dilated ascending colon up to hepatic flexure. Terminal ileum of normal caliber	Divided stoma of ascending and transverse colon
6	Ileocolic	II	Condition of bowel healthy	Excision proximal dilated ileum and 1 cm of distal atretic colon. End-to-oblique ileocolic anastomosis

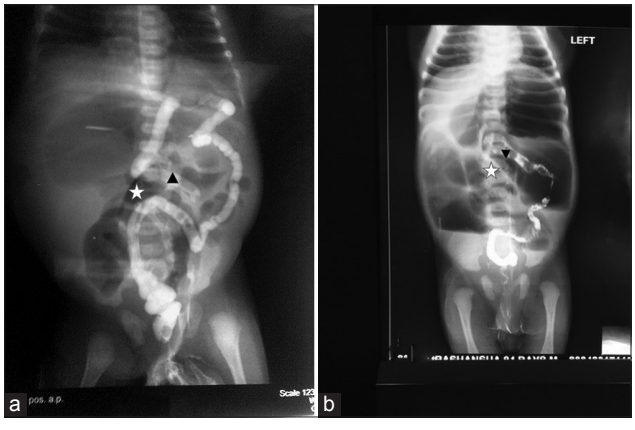

Figure 3: (a) The right half of transverse colon (black triangle) is bent back on itself to form a "Hook" by the adjacent ascending colon loop. Case of Type IIIa atresia at hepatic flexure (white star), (b) transverse colon is seen to turn back on itself (black triangle) to form a "hook or a hairpin shape" by adjacent dilated ileal loops (white star). Case of Type II ileocolic atresia

researchers have reported CA without mesenteric involvement in experimental animals, with defects in either fibroblast growth factor 10 or fibroblast growth factor receptor 2b [11]. However, these hypotheses cannot explain the genesis of CA in our case of sigmoid atresia with ARM (Case 4; Figure 1). In this patient, though the mesenteric and vascular defects ware near complete, the corresponding colonic defect was limited to a small portion of the mid-sigmoid region, the proximal colon segment was being supplied by a marginal artery. For the colon to be viable despite obliteration of its three main vessels, a slow sequential obliteration of the mesentery and its vasculature would provide more chance for the marginal artery to take over as the main arterial supply of the colon. Moreover, the anatomy of the colon vasculature suggests that the ascending branch of the left colic artery is the primary supplying vessel in 96.91% cases, and an intact left colic artery, including its collaterals at the splenic flexure, will supply sufficient blood to the proximal ascending colon after central ligation of the middle and right colic arteries [12]. We speculate that the sequence of obliteration could have been - ileocolic, right colic, middle colic, and finally, the left colic artery. The phenomenon could have occurred later in gestation, first involving the vessels supplying the distal midgut and later the proximal hindgut, sparing the marginal artery. Consequently, the marginal artery (continuation of ileal artery) became the sole supply to the colon and the mid-sigmoid colon became the farthest region in its perfusion territory. As the ileal artery could not have supplied this region adequately, it became atretic over time. The viability of the distal rectosigmoid colon can be attributed to its alternative supply by the inferior vesical and middle sacral arteries, and the arteries supplying the levator ani [13].

We could not assign a place to this defect within the existing classification. It did not have the features of a typical Type IIIa atresia, that is, a V-shaped mesenteric defect adjacent to the atretic bowel, and instead, there were some features similar to an "apple-peel" atresia such as a large mesenteric defect, an anastomotic arcade from the ileocolic vessel precariously supplying the distal segment and chronic ischemic damage, although the supply was antegrade rather than retrograde. Moreover, there was no spiraling of mesentery as seen in apple-peel deformity. As this is a first such reported presentation of CA, we admit that the above discussion is a speculation rather than a hypothesis.

Reported diagnostic signs on contrast enema suggesting CA include the "club" or "cobra head" sign, suggesting Type I CA and the "hook" sign suggesting Type III CA [14,15]. The "hook" sign describes a distal loop recurved on itself and lying juxtaposed to the gas-filled dilated proximal loop and is regarded by authors as being typical for Type III atresia [16]. This sign was present in three of our patients-Type IIIa atresia at hepatic flexure (n=2) Type II ileocolic atresia (n=1) (Figures 2 and 3). Thus, the sign is not specific for Type III atresia. Presumably, the meconium-filled, progressively enlarging proximal loop (ileal or colonic) pushes and displaces the relatively smaller and lighter microcolon which tends to fold on itself over time. Another striking finding seen on contrast enema in our series was that the colon failed to cross the midline to the right side and there was no cecal shadow (Figures 2 and 3). This seems more suggestive of CA.

The choice of surgical procedure, especially for right-sided CA is a matter of contention. A primary stoma is usually advocated in cases of a sick child, unhealthy bowel, major bowel discrepancy, complicated CA, or major associated anomalies [16]. Others recommend it due to their experience of an increased anastomotic dehiscence rate and mortality after a primary anastomosis [17]. However, a 100% success rate with primary anastomoses in uncomplicated cases has also been reported [8,18]. A worldwide survey in 2005 indicated that surgeons perform a staged management nearly thrice as frequently as a primary anastomosis [4]. Of late, primary anastomoses is preferred in contemporary practice in the absence of the above-mentioned factors [19]. We do not look for an associated HD in these cases. As such, HD is present in up to 2% cases of CA [19]. We agree with England et al. that this association is so rare that a primary anastomoses is justified without a rectal biopsy [20].

Nine of our babies had an isolated CA, a good birth- weight and all were discharged after the initial procedure. This is in agreement with the reported observation that isolated CA has a good outcome. However, 4/7 (57.1%) babies on stoma were readmitted with dehydration with one baby dying after the second admission despite our routine practice of prescribing ORS-zinc solution to all neonates with a stoma, to minimize chances of dehydration. This underscores the need and difficulties in managing high stoma output in neonates within the prevailing socioeconomic conditions of patients in developing countries. On the other hand, all three patients with a primary anastomosis survived and had a satisfactory postoperative outcome. We agree with Chadha et al. that to prevent morbidity associated with creation of stomas, a primary anastomosis should be performed wherever possible and if a stoma is fashioned, the patients should be kept on close follow-up with early closure of the stoma [8]. Although we encountered peristomal excoriation in nearly every case following stoma formation, there was no incidence of stomal prolapse in these cases.

The mortality rate in our series was 20% (2/10) comparable to the reported 28% overall mortality in CA [4]. Others have reported mortality figures varying from 0% to 61% [17]. Other factors reportedly responsible for a higher mortality are prematurity, time to operation more than 72 h after birth, multiple atresia's, associated anomalies, a missed atresia, and anastomotic leak [4,6,16,17].

CONCLUSION

Although an acute antenatal vascular accident is probably the most important etiological factor, CA can also result from a gradual, sequential obliteration of the mesenteric vasculature. A primary anastomosis should be contemplated if feasible as high stoma output, consequent dehydration leads to high morbidity and mortality, especially in developing countries. The high stoma output should be actively compensated with use of ORS zinc solution. A close follow-up and an early stoma closure should be planned.

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