Different varieties of colonic atresia in a series of 13 patients: a single-center experience

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Purpose **The aim of this study was to review our experience in the management and outcome of colonic atresia, either isolated or associated with anorectal anomalies.**

Methods **A total of 13 neonates with colonic atresia were operated upon in the Pediatric Surgery Unit of Zagazig University Hospital between August 2004 and April 2010. Complete data including age, sex, clinical findings, diagnostic procedure, associated anomalies, preoperative management, intraoperative findings, operative procedures, postoperative course, and outcome were reported for each patient.**

Results **Colonic atresia was found in 13 patients, including nine boys (69.2%) and four girls (30.8%). It was located in the ascending colon in four (30.8%) patients, in the transverse colon in three (23%), and in the sigmoid colon in four (30.8%). The other two (15.4%) patients had colonic atresias extending to the small bowel. Two patients had associated anorectal anomalies. Type III colonic atresia was observed in six patients (46.2%), type II in three (23%), and type I in four patients (30.8%). The treatment included resection and primary anastomosis in seven patients (53.8%) and diverting colostomy in six (46.2%). Two patients died: one because of severe neonatal sepsis and the other because of severe cardiac anomalies. One patient developed anastomotic leakage and another had anastomotic dysfunction.**

Conclusion **Colonic atresia is an uncommon cause of neonatal intestinal obstruction. The combination of colonic atresia and anorectal anomalies should be taken into consideration. Proper postoperative care and management of postoperative complications improve the outcome.** ***Ann Pediatr Surg*** **9:20–24 © 2013 Annals of Pediatric Surgery.**

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Introduction

Colonic atresia is a very rare cause of intestinal obstruction and constitutes ~1.8–15% of all intestinal atresia cases. Limited data are available on its management and outcome [1]. Up to 75% of all colonic atresias are found proximal to the splenic flexure, usually in the ascending colon [2].

Neonates with colonic atresias show typical clinical features of low intestinal obstruction with abdominal distention, bilious vomiting, and failure to pass meconium [3]. Plain radiographs are usually nonspecific and show features of low intestinal obstruction in the form of dilated bowel loops, multiple air-fluid levels, and absence of air in the rectum. The diagnosis is confirmed by contrast enema, which is the study of choice. The part of the colon distal to the atresia shows a microcolon with failure of retrograde flow of contrast [4].

The management and outcome of colonic atresia have improved in the last few decades because of several factors, including advances in neonatal anesthesia, neonatal intensive care, and operative technique, and use of total parenteral nutrition [5].

The objective of this study was to review our experience in the management of colonic atresia, either isolated or associated with anorectal anomalies, with regard to clinical presentation, diagnostic modalities, intraoperative findings, choice of procedure, postoperative management, and outcome of the patients.

Materials and methods

A total of 13 neonates with colonic atresias were operated upon in the Pediatric Surgery Unit of Zagazig University Hospital between August 2004 and April 2010. Complete data including gestational age, age on presentation, sex, clinical findings, diagnostic procedure, associated anomalies, preoperative management, intraoperative findings (site and type of atresia, associated gastrointestinal anomalies, and operative procedures), postoperative course, and outcome were reported for each patient.

The 13 patients were classified into four types according to Grosfeld's classification [6]: a type I defect represents a mucosal defect with an intact mesentery; type II consists of a fibrous cord connecting the atretic bowel ends; a type III lesion denotes an atretic segment with a V-shaped mesenteric gap defect; and type IV describes instances of multiple atresias.

At the time of admission, a nasogastric tube was placed for gastric decompression. Complete laboratory investigations were performed for blood profile, blood gases, electrolytes, coagulation profile, and liver and kidney function, if indicated. Fluid and electrolyte imbalances were corrected. Plain abdominal radiography was performed if the findings were suggestive of large bowel obstruction; subsequent contrast enema was performed on the same day using water-soluble nonionic contrast.

As a routine, preoperative antibiotics cephalosporin and metronidazole were administered. The time between

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presentation and surgery varied from 12 h to 3 days. The delay was due to preoperative attempts at resuscitation and lack of space in the neonatal intensive care unit in some cases.

During laparotomy, complete abdominal exploration was undertaken to detect the site and type of colonic atresia and associated gastrointestinal anomalies such as small bowel atresia, malrotation, and/or volvulus. Patency of the distal bowel was ensured by injection of saline. The operative technique varied from resection and single-layer anastomosis using 5/0 vicryl interrupted sutures for right-side colonic atresia to colostomy with mucous fistula for left-side colonic atresia or, in the case of critically ill patients, to avoid prolonged operation. Rectal biopsy was taken during the operation for histopathological examination to exclude the presence of associated Hirschsprung's disease (HD).

Nasogastric tube aspiration, IV fluid, and antibiotics were continued postoperatively. Oral feeding was started at least on the fourth postoperative day in the case of primary anastomosis if the patient passed stool and the nasogastric tube aspirate was clear and less than 30 ml/24 h. Low-volume milk, either expressed breast milk or milk formula, was given through the nasogastric feeding tube, and, if the patient tolerated, the volume was gradually increased until total enteral nutrition could be instituted.

Patients were discharged from the hospital when full enteral nutrition was reached. The length of post-operative hospital stay ranged from 7 days to 1 month. The patients were followed up in the outpatient clinic every week during the first month and then every month for 1 year to ensure the growth of the child and absence of obstructive symptoms. Patients with colostomy were readmitted at our hospital 2 months later for closure of colostomy or for performing definite repair on the patients with associated anal anomalies.

Results

Thirteen neonates were found to have colonic atresia, including nine boys (69.2%) and four girls (30.8%). Age at the time of presentation ranged from 1 to 5 days. All patients presented with abdominal distension, bilious vomiting, and failure to pass meconium. Plain abdominal radiographs showed a dilated bowel loop with air-fluid levels. Water-soluble contrast enemas were performed for all patients except two who had associated anorectal anomalies and showed a distal microcolon with complete cut-off of the colon contrast column and for one patient who showed a rounded 'cobra-head' deformity (Fig. 1).

During the operation, the site and type of colonic atresia were determined (Table 1). Colonic atresia was located in the right colon (cecum, ascending colon, and hepatic flexure) in four (30.8%) patients, in the transverse colon in three (23%), and in the sigmoid colon in four (30.8%) patients. The other two (15.4%) patients had colonic atresia extending to the small bowel: one had ileocecal atresia extending from the terminal ileum to the cecum and the other had jejunocolic atresia extending from the jejunum 20 cm distal to the ligament of Treitz to the mid-transverse colon. Three types of colon atresias were found: type III was observed in six (46.2%) patients, type II in three (23%), and type I in four (30.8%).

Fig. 1

Contrast enema demonstrated cobra head deformity in a case of membranous atresia between the cecum and the ascending colon.

The associated anomalies are presented in Table 1. Two neonatal patients were found to have colonic atresia and associated anorectal anomalies: one had sigmoid atresia and anorectal atresia and the other had sigmoid atresia, rectal atresia, and anovaginal fistula (Fig. 2).

During primary surgery, resection and primary anastomosis were performed in seven (53.8%) patients and diverting colostomy in six (46.2%) (Table 2). Four patients were treated using segmental ileocolic resection and primary end-to-oblique anastomosis, one using jejunocolic anastomosis and tapering jejunal enteroplasty, two using primary colocolic anastomosis, two using transverse colostomies, and four using left-side colostomies.

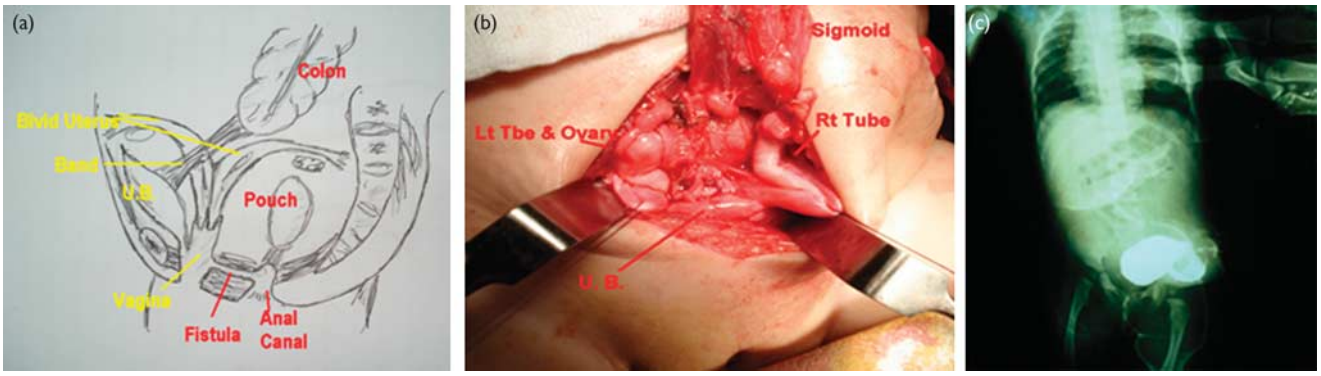
Postoperative complications were recorded (Table 2). Two patients died; the cause of death was cardiac anomalies in case 9 and neonatal sepsis in case 11. One patient developed anastomotic leakage and a diverting stoma was created. One patient had anastomotic dysfunction and did not tolerate oral feeding; total parenteral nutrition was given for 2 weeks and then oral feeding was restored with a gradual increase in volume until full enteral nutrition was reached. During the follow-up period at the outpatient clinic, four patients with colostomy were readmitted: two for closure of colostomy and the other two for undergoing posterior sagittal anorectoplasty and colonic pull-through. The postoperative course was smooth for these patients and no

Table 1 Clinical and intraoperative data

Case number	Age (days) and sex	Site of atresia	Type of atresia	Associated anomalies
1	2, M	Transverse colon to splenic flexure	Type III	Limb anomalies
2	1, M	Ascending colon	Type II	–
3	4, M	Sigmoid colon	Type I	Malrotation and Meckel's diverticulum
4	1, M	Ascending colon	Type III	–
5	1, F	Sigmoid colon	Type I	–
6	2, M	Transaverse colon	Type I	Malrotation
7	1, F	Ascending colon and hepatic flexure	Type III	Unilateral cleft lip
8	3, M	Transverse colon	Type II	–
9	2, M	Sigmoid colon	Type II	Anorectal atresia, facial anomalies, limb anomalies, cardiac anomalies
10	1, M	Ileocecal atresia	Type III	Ileal atresia
11	3, F	Jejuno-transverse colon	Type III	Jejunal atresia
12	5, M	Ascending colon	Type I	–
13	1, F	Sigmoid colon	Type III	Rectal atresia, bifid uterus and anovaginal fistula

F, female; M, male.

Fig. 2



(a, b) The diagram and the image show sigmoid atresia associated with rectal atresia, bifid uterus, and anovaginal fistula. (c) A distal loopogram depicts sigmoid atresia in the same patient.

Table 2 Management and outcome

Case number	Surgical techniques	Early complications	Secondary operation	Outcome
1	Double-barrel colostomy	–	Closure of colostomy	Alive
2	Colocolic anastomosis	Anastomotic dysfunction	–	Alive
3	Double-barrel colostomy	–	Closure of clostomy	Alive
4	Ileocolic anastomosis	–	–	Alive
5	Double-barrel colostomy	–	–	Loss to follow-up
6	Ileocolic anastomosis	Leakage	Diverting stoma	Alive
7	Colocolic anastomosis	–	–	Alive
8	Double-barrel colostomy	–	Closure of clostomy	Alive
9	Double-barrel colostomy	–	–	Died
10	Ileocolic anastomosis	–	–	Loss to follow-up
11	Jejuno-transverse anastomosis and tapering jejunoplasty	Prolonged ileus and sepsis	–	Died
12	Ileocolic anastomosis	–	–	Alive
13	Double-barrel colostomy	–	Combined abdominal and PSARP, rectal mucosectomy, colonic pull-through	Alive

PSARP, posterior sagittal anorectoplasty.

complications were recorded. Two patients were lost during the follow-up period.

Discussion

The common theories explaining the cause of colonic atresia are a lack of revacuolization of the solid cord stage of intestinal development and a late intrauterine mesenteric vascular accident [7].

Grosfeld's classification has been applied to colonic atresia [6]. Type III was the most frequent type in previous reports (80%), followed by type I and type II [8]. However, in this study, type III atresia was present

in 46.2% of patients, whereas type I and type II were present in 30.8 and 23%, respectively. The sites of type III colonic atresia were similar to those reported in the literature [2,8,9]. They were usually proximal to the splenic flexure.

The association of colonic atresia with other congenital anomalies is rare [10]. However, two patients with malrotation were encountered in this study. Meckel's diverticulum, anal atresia, limb anomalies, and cleft lip were also observed.

The presence of colonic atresia associated with anorectal anomalies is extremely rare and only five cases have been reported in the English literature [11]. In this study, two patients with sigmoid atresia and associated anorectal anomalies were reported: one was associated with anorectal agenesis and the other with rectal atresia and anovaginal fistula. The conditions seen in the latter patient have not been previously reported in the literature.

A significant feature in this research was the delayed presentation of several patients. The occurrence of preoperative hypovolemia, dehydration, electrolyte imbalance, and sepsis has an adverse effect on the prognosis. Late referral of patients and lack of facilities at the neonatal intensive care unit were also adverse prognostic factors in this research. One of the most important factors was that surgery had to be performed either by or under the supervision of an experienced pediatric surgeon.

In the past decades, a staged approach for management of colonic atresia was preferred, aiming to avoid hazards of bowel anastomosis [5,8]. The recommended procedure was to perform resection and primary anastomosis for colonic atresia proximal to splenic flexure and to perform a colostomy for more distal colonic atresia [12]. This procedure was also recommended by Benson *et al.* [13]. Both staged management and primary anastomosis were used by Pohlson *et al.* [9]. Several authors have reported the use of resection and primary anastomosis as a reasonable treatment option regardless of the location of the colonic atresia [14].

In this study, we preferred to perform resection and primary end-to-oblique anastomosis on the patients with right colonic atresias and to perform double-barrel colostomy for distal atresias or in unstable patients to avoid the hazards of prolonged operation. In one patient with severe atresia extending from the jejunum to the mid-transverse colon, tapering enteroplasty was performed in addition to primary anastomosis to restore bowel caliber and save intestinal length.

The combination of colonic atresia and HD is rare and the diagnosis of HD is usually delayed after the surgical treatment of colonic atresia. When complications occur, the morbidity and mortality of patients increase [15]. For this reason, routine rectal biopsy was taken to exclude the presence of HD. In this study, a combination of colonic atresia and HD was not reported.

Cox *et al.* [16] operated on six of 14 patients with colon atresia with primary colonic anastomosis. Two of them developed complications because of unrecognized distal hypoganglionosis. Levard and Boureau [17] reported seven cases of congenital colonic atresias over a 13-year period. Two patients had postoperative complications: one had an anastomotic leak and one had a dysfunction, which were successfully treated by other operations. Chadha *et al.* [18] mentioned that prognosis was good in three of the four patients surviving resection and primary anastomosis for right-sided colonic atresia.

In the current study, one patient developed anastomotic leakage and a stoma was created. Another patient developed anastomotic dysfunction and was given TPN. This patient improved with conservative management and did not need a secondary operation.

The mortality of colonic atresia varies from 0 to 50% and outcomes differ because of various factors [14]. Complications such as dehydration, electrolyte imbalance, aspiration of vomitus, sepsis, and malnutrition are also proposed causes of death [8]. The death of two patients (17.2%) reported in this study was because of severe neonatal sepsis and cardiac anomalies.

Conclusion

Colonic atresia is an uncommon cause of neonatal intestinal obstruction. The association between anorectal anomalies and colonic atresia should be taken into consideration. Selection of the operative technique depends on the location of the atresia and the preoperative general condition of the neonates. Resection and primary anastomosis are preferred for right-side colonic atresia, whereas double-barrel colostomy is preferred for left-side colonic atresia. Proper postoperative care and early recongition of postoperative complications and management improve the outcome of the cases.

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Conflicts of interest

There are no conflicts of interest.

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