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Colonic atresia: surgical management and outcome

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Abstract Colonic atresia (CA) is a very rare cause of intestinal obstruction, and little information has been available about the management and predictors of outcome. A retrospective clinical trial was performed to delineate the clinical characteristics of CA with special emphasis on surgical treatment and factors affecting outcome. Children with CA who were treated in our department between 1977 and 1998 were reviewed: 14 boys and 4 girls aged 1 day to 5 months. All but 2 referred patients and 1 with prenatal diagnosis presented with intestinal obstruction. Plain abdominal X-ray films showed findings of intestinal obstruction in 14 cases; a barium enema demonstrated a distal atretic segment and microcolon in 4. The types of atresia were IIIa (n = 9), I (n = 6), and II (n = 3). Type IIIa atresias were located proximal to the splenic flexure (n = 8) and in the sigmoid colon (n = 1), type I atresias were encountered throughout the colon; and all type II atresias were proximal to the hepatic flexure. Associated anomalies were multiple small-intestinal atresias (MSIA) (n = 4), gastroschisis (GS) (n = 2), pyloric atresia (n = 1), Hirschsprung's disease (n = 1), and complex urologic abnormalities (n = 1). The initial management was an enterostomy in 15 patients (83%), including 2 referred and 2 with GS, and primary anastomosis in the remaining 3 (17%). Secondary procedures were the Santulli operation (n = 2), colostomy closure and recolostomy followed by a Swenson operation (n = 1), sacroabdominoperineal pull-through (n = 1), and colostomy closure (n = 1). Leakage was encountered in all primarily anastomosed patients. The overall mortality was 61%. Deaths occurred in patients with associated major anomalies (GS 2, MSIA 3, pyloric atresia 1) (55%) and in 3 patients who were initially managed by primary anastomosis (27%). Two additional patients died of sudden infant death syndrome (18%). Type I atresia was more common than in previously reported series and was associated with proximal multiple atresias. The initial management of CA should be prompt decompression of the intestine by an ostomy procedure, preferably end- or double-barrel. The type of surgery (primary anastomosis without prior colostomy) and associated abnormalities are the major determinants of poor outcome.

Keywords Atresia · Colon · Malformation · Management · Newborn

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Introduction

Colonic atresia (CA) is an extremely rare cause of intestinal obstruction and constitutes approximately 1.8% to 15% of all intestinal atresias [14]. Because of its rarity, little information has been available about the management and predictors of outcome. Therefore, a retrospective clinical trial was performed to delineate the clinical characteristics of CA with particular emphasis on surgical treatment and factors affecting outcome.

Patients and methods

The records of infants with CA who were treated at the Department of Pediatric Surgery of Hacettepe University Medical Faculty between 1977 and 1998 inclusive were retrospectively reviewed. Information recorded for each patient included age, sex, presenting symptoms and signs, diagnostic procedures, location and type of atresia, associated abnormalities, surgical treatment, histopathologic findings, and outcome. The type of atresia was determined according to Grosfeld's classification in which type I represents mucosal atresia (membranous) with intact bowel wall and mesentery, type II blind ends separated by a fibrous cord, type IIIa blind ends separated by a V-shaped mesenteric defect, type IIIb apple-peel atresia, and type IV multiple atresias [7].

Results

Eighteen children (14 [78%] boys and 4 girls [22%]) were treated for CA during the study period. The clinical data are summarized in Table 1. The age at presentation ranged from 1 to 5 days in 16 patients who were primarily managed in our hospital. Two patients aged 3 and 6 months were initially managed in other hospitals. All but 1 were term babies born by vaginal delivery with normal birth weights. One was delivered by cesarean section because of preterm labor.

Most of the patients presented with failure to pass meconium, abdominal distention, and bilious vomiting. Two patients with gastroschisis (GS) were admitted due to the abdominal defect, and the accompanying CA was diagnosed at the initial examination. Two patients were referred to our unit after colostomies were performed. Plain abdominal X-ray films revealed air-fluid levels (n = 14), pneumoperitoneum (n = 1), calcification (n = 1), and isolated stomach air (n = 1). Barium enemas were not in routine use until 1985. Therefore, the diagnosis of CA was made by barium enema only in 4 patients.

The types of atresia were IIIa in 9, I in 6, and II in 3 (Table 1). Multiple type I atresias and type I atresia accompanying type IIIa atresia were also encountered in 2 patients. Type IIIa atresias were located proximal to the splenic flexure in either the ascending or transverse colon (n = 8) and in the sigmoid (n = 1). Type I atresias (n = 6) were encountered in all locations from the cecum to the sigmoid. All type II atresias (n = 3) were proximal to the hepatic flexure. There was a significant amount of colon missing between the blind atretic segments in all type IIIa cases. In type II cases the distance between the blind atretic segments was a few centimeters in 2 and most of the colon was absent in the remaining patient.

Associated anomalies are shown in Table 1. The most common was multiple small intestinal atresias (MSIA) (n = 4), which were found in association with types I (n = 3) and IIIa (n = 1) CA. GS (n = 2), Meckel's diverticulum (n = 2), pyloric atresia (n = 1), crossed renal ectopia and vesicoureteral reflux (n = 1), malrotation (n = 1), atypical face with minor extremity anomalies (n = 1), Hirschsprung's disease (HD) (n = 1), and omphalocele (n = 1) were the other anomalies.

Sixteen cases were initially managed in our unit; 13 of them initially underwent ostomy procedures, either end- (n = 5) or double-barrel colostomies (n = 5), the Santulli operation (n = 1), ileostomy and pyloroplasty (n = 1), and a modified Lester Martin operation (n = 1) (Table 2). An ileocolic anastomosis was the mode of treatment in the remaining 3. Patients with GS underwent primary or patch repair of the abdominal defect and colon exteriorization.

Bowel continuity was established by Santulli operation in 2 cases, a Swenson operation in 1 (in a patient with HD), sacroabdominoperineal pull-through in 1, and colostomy closure in 1, in patients who were initially treated by bowel exteriorization. Pathologic examination of the resected specimens did not reveal any special histopathologic diagnosis apart from the 1 patient with HD.

Anastomotic leakage was the most common postoperative complication (n = 4) in our series. Iatrogenic injuries were jejunal perforation (n = 1) and urethral transection (n = 1). Sepsis in 2 and respiratory distress in both GS patients were also noted in the early postoperative period.

The overall mortality was 61% in our series. Most occurred in patients with associated major anomalies (GS 2, MSIA 3, pyloric atresia 1) and in patients initially managed by primary anastomosis (n = 3). Two

Table 1 Epidemiologic and clinical characteristics

Case no.	Age (days) and sex	Location of atresia	Type of atresia	Associated anomalies
1	3, F	Mid-transverse to splenic flexure	Type IIIa	–
2	6 months, M	Sigmoid colon	Type IIIa	Cross renal ectopia and vesicoureteral reflux
3	4, M	Hepatic flexure to splenic flexure	Type IIIa	Hirschsprung's disease, Meckel's diverticulum
4	3, F	Mid-transverse	Type I	Malrotation, atypical face, minor abnormalities in lower extremities
5	2, F	Hepatic flexure	Type I	–
6	4, M	Sigmoid colon	Type I	Type IIIa and multiple type I atresias in small intestine
7	5, F	Ascending colon and rectum	Type IIIa + type I	–
8	3, M	Ascending and transverse colon	Multiple type I	–
9	3 months, M	Ascending to splenic flexure	Type IIIa	Omphalocele
10	5, M	Cecum	Type II	–
11	2, M	Cecum	Type II	–
12	3, M	Hepatic flexure to splenic flexure	Type IIIa	–
13	1, M	Ascending colon to rectum	Type II	Type I pyloric atresia and Meckel's diverticulum
14	3, M	Transverse colon	Type I	Multiple type I and II jejunoileal atresias
15	4, M	Cecum	Type I	Multiple type I ileal atresias
16	1, M	Cecum to sigmoid colon	Type IIIa	Gastroschisis
17	1, M	Transverse colon	Type IIIa	Gastroschisis
18	1, M	Transverse colon	Type IIIa	Multiple type I jejunoileal atresias

Table 2 Management and outcome

Case no.	Initial operation	Secondary operation	Complication	Result and outcome
1	End-colostomy	Santulli operation	–	Sudden infant death
2	End-colostomy[a]	Sacroabdominoperineal pullthrough	Transection of urethra	Sudden infant death
3	End-colostomy	Anastomosis, afterward recolostomy, Swenson op.[b]	Anastomotic leak	Alive
4	Double-barrel colostomy	–	–	Lost to follow-up
5	End-colostomy	–	–	Lost to follow-up
6	Modified Lester Martin operation and sigmoid end-colostomy	Repair of leakage and gastroenterostomy	Leaks	Died
7	Ileocolic anastomosis and rectal web perforation	–	Sepsis, meningitis	Died
8	End-colostomy	–	–	Loss of follow-up
9[a]	End-colostomy	Santulli op., afterward colostomy closure	–	Alive
10	Ileocolic anastomosis	Double barrel ostomy	Closed perforation	Died due to sepsis
11	Santulli operation	–	–	Alive
12	Double-barrel colostomy	–	Sepsis	Died due to sepsis
13	Ileostomy and pyloroplasty	–	Iatrogenic jejunal perforations	Died
14	Colonic web excision and small intestinal resection with anastomosis	–	Leak and sepsis	Died
15	Resection of atretic ileal segment and double-barrel ostomy	–	–	Died
16	Graft repair of gastroschisis and double-barrel colostomy	–	Respiratory distress	Died
17	Primary gastroschisis closure and end-colostomy	–	Respiratory distress	Died
18	End colostomy and resection of small intestinal atresias and anastomosis	Colostomy closure	–	Alive

[a] Omphalocele repair and end-colostomy were performed in another hospital
[b] Rectal biopsy confirmed diagnosis of Hirschsprung's disease and patient underwent Swenson's operation

additional patients died from sudden infant death syndrome (SIDS) in the 1st and 2nd postoperative months.

Discussion

A vascular insult of the fetal intestine due to volvulus, intussusception, an internal hernia, or strangulation in a tight GS or omphalocele defect is universally accepted to be the major cause of all types of CA [11, 19]. Familial instances of CA have been observed, suggesting that genetics may play a role in some cases [8, 18]. A male predominance was encountered in our series, similar to other previous reports [16, 21].

The currently used classification of intestinal atresias was proposed by Grosfeld and has also been applied to CA [7, 12]. Type IIIa has been the most frequent type of CA in previous reports (80%), followed by types I and II [3, 16, 21]. However, in our series type IIIa atresia was only noted in 50% of cases. Type I atresia was more common in our series (33%) than in previous reports; furthermore, most of the type I atresias were associated with more proximal multiple atresias that required more extensive operations, complicating the postoperative course. The locations of the type IIIa atresias were similar to those reported in the literature [3, 16, 21]. They were usually proximal to the splenic flexure except 1 in the sigmoid colon. On the other hand, type I atresias occurred throughout the colon. The variable locations of type I atresias and frequent association with MSIA necessitates a careful search of the whole intestinal tract. Because associated type I atresias may not be recognized on macroscopic examination, the patency of the intestinal tract should be checked by instillation of physiologic saline. The ratio and location of type II atresias showed similarity to previously reported cases [3, 21].

Infants with CA show the typical clinical features of low intestinal obstruction with distention, vomiting, and failure to pass meconium. The vomitus is usually bile-stained. It may be clear initially and may not be bile-stained in patients with associated obstruction proximal to the ampulla of Vater. In infants with associated GS, the diagnosis is usually made on inspection. Although prenatal ultrasonographic (US) examination may yield a suggestive diagnosis of intestinal obstruction, its ability to exclude a colonic malformation is limited. However, CA has been reported to be diagnosed by US in the early hours of life before the occurrence of distention and radiologic findings [15].

Plain abdominal X-ray films show multiple loops of distended intestine with air-fluid levels, and there is often a hugely distended segment corresponding to the dilated proximal colon. A barium enema demonstrates the location of the distal atretic segment. Although these studies were not obtained in the early years of our series, we strongly recommend them in all types of neonatal

intestinal obstruction. In other words, a plain radiograph suggesting a proximal obstruction does not make a barium enema unnecessary. Conversely, a barium enema demonstrating CA does not exclude the possibility of associated more proximal atresias in either the colon or small intestine. This is obvious from our series based on the high incidence of associated proximal atresias. A barium enema also shows meconium pellets in instances of meconium ileus and a transitional zone in HD.

Associated anomalies are reported to be rare [11]. However, it appears that GS and MSIA are common, occurring in 10% and 22% of patients, respectively. Complex urologic abnormalities were encountered in 1 patient. An atypical appearance of the face was also noted in previous reports with similar incidence (6%) [3, 16]. HD, a rare association with CA, has also been reported in the literature and is usually suspected following disruption of the anastomosis of the atretic colon segments [1, 10].

Operative management of CA has been refined over the years. In earlier reports, a staged approach was recommended on the basic concept of initial prompt relief of obstruction and to avoid the hazards of primary anastomosis. After a few weeks or months, when the infant is thriving, the enterostomy is taken down and intestinal continuity is established by an appropriate end-to-end or end-to-side anastomosis [3, 21]. Santulli and Blanc recommended a technique providing rapid decompression of the intestine without requiring an additional operation for stoma closure [19]. The major handicap of this procedure is creating an intra-abdominal anastomosis. Therefore, it should not be used in complicated cases. Initial decompression can also be accomplished by double-barrel colostomy.

Hartman et al. recommended that a double-barrel colostomy should be the primary operation, with closure deferred for 9–12 months [9]. Sturim and Ternberg preferred resection of the proximal colon and a primary ileocolic anastomosis in atresias proximal to the splenic flexure, and an initial colostomy followed by subsequent resection and anastomosis for more distal atresias [20]. This policy was also recommended by Benson et al. and Coran and Eraklis [2, 4]. Freeman preferred resection of the proximal dilated colon and primary anastomosis [5]. Both staged management and primary anastomosis have been used by Pohlson et al. [16].

We preferred end-colostomies as the initial approach in the earlier years of our series. The Santulli operation was also used, both as an initial or secondary operation, in some cases. Primary anastomosis following resection of the proximal dilated segment was used in some cases as an initial approach and a modified Lester Martin operation was performed in 1. All patients who were initially managed by primary anastomosis died due to anastomotic leakage causing sepsis. Thus, primary repair cannot be recommended in light of our results.

In cases with associated GS the problem is somewhat more complex, but the same principles can be applied. Initial end-colostomy with repair of the GS is recommended by some authors [3, 6, 17]. The major reasons for creation of the ostomy were bowel damage from amniotic exposure and preatretic dilatation. Additionally, attempts at primary anastomosis resulted in anastomotic stenosis and/or leakage [6]. HD is an important pathology in patients with CA. The suspicion of this pathology should arise in the presence of anastomotic leakage [1, 10]. This course was also seen in 1 of our cases, and treatment of the megacolon obviated the problem.

The mortality of CA varies between 0% and 50%, and the outcome differs due to various factors [7, 13, 16, 19]. In earlier years, the anesthetic and operative shock were held responsible for most of the early postoperative deaths. Increased mortality has been reported in patients with colonic obstruction diagnosed after 4 days of age [20]. The complications of dehydration, electrolyte imbalance, aspiration of vomitus, peritonitis, infection, malnutrition, and postoperative adhesions have been also proposed as causes of death. In addition, in light of the present study, we conclude that whenever the lesion is treated by primary anastomosis, the expected result is nonfunction or leakage of the anastomosis. The presence of associated MSIA and GS affected survival adversely. On the other hand, the mortality due to SIDS following a reasonable period after the surgery emphasizes the importance of close observation of these patients after discharge.

In conclusion, we emphasize that type I CA was more common than in previously reported series and associated with multiple proximal atresias. The initial management of CA should be prompt decompression of the intestine by an ostomy procedure, preferably end- or double-barrel. The type of surgery (primary anastomosis without prior colostomy) and associated abnormalities are the major determinants of poor outcome.

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