Colonic Atresia

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Contents

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73.1 Introduction

Atresia of the colon is an uncommon entity distinct from congenital pouch colon, which is a more frequent occurrence in India and Asia and is associated with anorectal malformations. Although the underlying cause may be vascular insufficiency [1, 2], the association with particularly Hirschsprung's disease [3–5] and the gross discrepancy between the proximal and distal bowel diameters militate against management strategies described for small bowel atresias.

73.2 Demographics

Atresia of the colon is a relatively rare form of intestinal atresia with an incidence of 1:40,000 to 1:60,000 live births and comprises less than 5% of the total number of gastrointestinal tract atresias (4.4% in our series of 248 bowel atresias distal to the duodenum) [6, 7]. There is no gender or racial predilection. There is an association with Hirschsprung's disease, gastroschisis, malrotation/nonfixation of the bowel, and a more proximal atresia of the small bowel [6]. The association with cloacal exstrophy is well known but will not be described here.

73.3 Etiology/Pathophysiology

The etiology of colonic atresia remains a subject of debate. The association with gastroschisis and intestinal atresia is thought to be as a result of a vascular accident in utero [2, 8, 9]. Other insults affecting vascular supply such as volvulus and constriction of bowel in a closing umbilical ring have been suggested [10].

It is not clear why aganglionosis is found in some, but it is thought that either the dilated loop proximal to the transition to aganglionosis undergoes volvulus with bowel ischemic injury and the development of atresia or it may suggest an early event whereby migrating nerve cells are arrested in their progress to populate the distal gut [4, 5]. This implies an early gestational interruption. An unfixed mesentery is often noted in cases of colonic atresia coexistent with Hirschsprung's disease suggesting intrauterine volvulus as a possible etiology [11, 12].

Colonic atresia can occur in any part of the colon with different case series showing predominance of the right or left colon. All types of atresia have been reported with type IIIa being more common in most series [6, 10, 13].

73.4 Clinical Presentation

73.4.1 History

Antenatal history may include that of polyhydramnios. Prenatal ultrasound may show a single dilated loop of bowel. Patients present with neonatal intestinal obstruction with gross abdominal distension if it is an isolated colonic atresia.

73.4.2 Physical

Aspiration of gastric content of a volume > 25 ml via orogastric or nasogastric tube or emesis which is bile stained along with abdominal distension and failure to pass normal meconium is suggestive of a distal bowel obstruction. Abdominal distension may not be so marked in cases with coexistent proximal jejunal atresia, a not infrequent association.

73.4.3 Investigations

The abdominal X-ray in isolated colonic atresia is typical in showing one or two very dilated gas and fluid-filled loops [13] (Fig. 73.1).

A pitfall to be avoided is the colonic atresia distal to a small bowel atresia. At laparotomy, the colon may look normal in outward appearance. It is, therefore, essential for the patency of the lumen of the colon to be evaluated in any small bowel atresia preferably preoperatively by contrast enema.

In addition, before the surgeon intends restoring colonic continuity, a preoperative contrast enema, which typically shows collapsed and disused distal colon (Fig. 73.2), needs to be performed to confirm patency of the distal bowel.

Biopsy of the bowel distal to the atresia showing normal innervation of the bowel to exclude Hirschsprung's disease is mandatory, if primary anastomosis is contemplated [13]. Likewise, rectal suction biopsy is done prior to stoma closure.

73.4.4 Management

Management should include full investigation for associated anomalies, exclusion of Hirschsprung's disease by biopsy of the bowel distal to the atresia [6], and exclusion of a second atresia or stenosis by contrast enema [14].

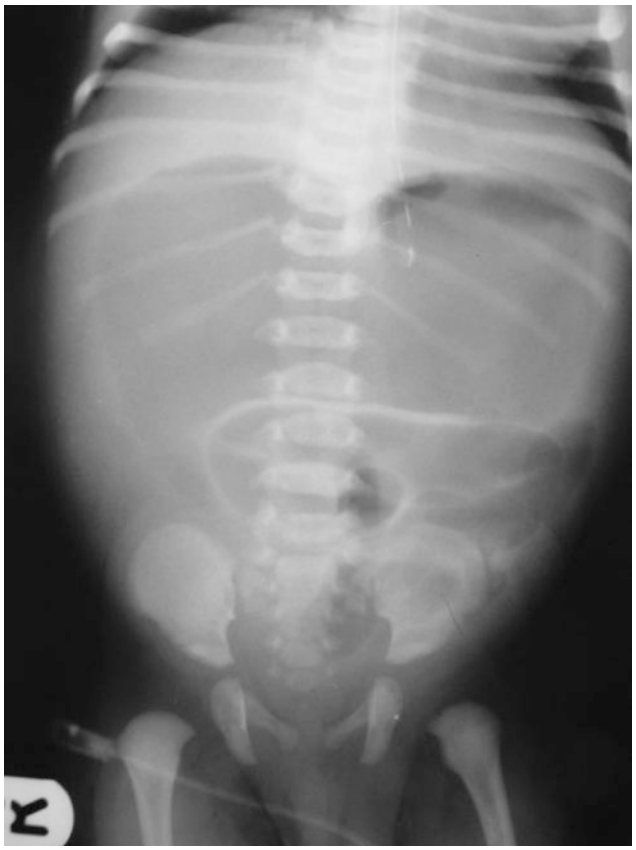

Fig. 73.1 AP abdominal radiograph showing a disproportionately large loop of bowel

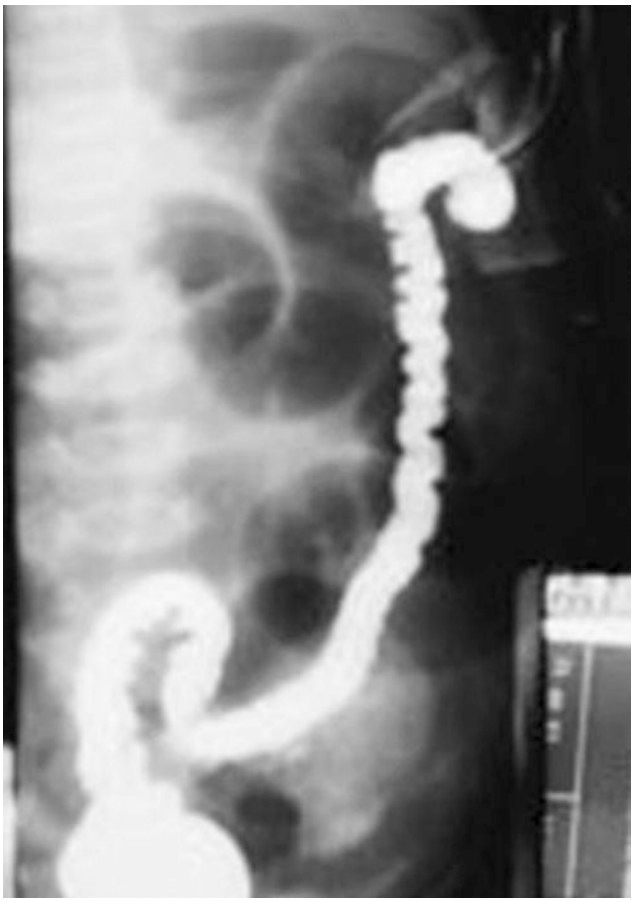

Fig. 73.2 Contrast enema of a patient with ileal atresia showing a sigmoid atresia and distal microcolon

The appropriate management in terms of best operative strategy remains a debate, possibly because of small case series and limited institutional experience on which to base management options. There has been little consistency of methods in many of the reported series [15]. The fashioning of a stoma at the level of the atresia is the preferred initial treatment rather than an attempt at primary anastomosis – especially if distal patency has not been confirmed or there is a large discrepancy in proximal and distal bowel diameter (Fig. 73.3). The gross dilatation of the blind end may need to be tapered to assist in fashioning a manageable stoma. This can then be followed with stoma closure once the baby is stable and thriving, and results of investigations on the distal bowel are determined.

As with small bowel atresias, both proximal and distal colonic ends adjacent to the atresia may have abnormal innervation and vascularity, and hence, at the time of anastomosis, a portion of the proximal dilated and distal micro-colon should be resected before anastomosis [16].

Primary resection and anastomosis can only be performed in selected cases if the distal colon is normal and bowel patency has been established, depending on coexistent pathology [16–18]. Primary resection and anastomo-

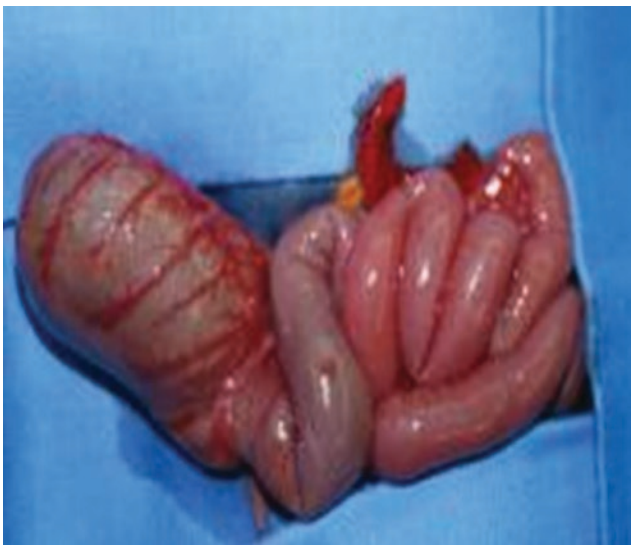

Fig. 73.3 Operative photograph of patient with colonic atresia at the hepatic flexure showing dilated ascending colon and collapsed transverse colon

sis have had higher complication rates in some series, usually due to undiagnosed distal pathology [13, 19], while a better prognosis is reflected in other series where most cases have been managed with a primary stoma [14].

73.5 Postoperative Complications

Missed diagnosis should not occur if the above principles are followed. In our series of 14 cases, there was one mortality due to a second colonic atresia, which was missed in a patient presenting with a small bowel atresia, causing a leak from the more proximal anastomosis, peritonitis, and death [13].

73.6 Prognosis and Outcomes

Outcomes should be good but are limited by associated intestinal atresia with short gut and the extent of other anomalies. Mortality in various series ranges from 7% to 61% [14, 19]. The type of surgery and coexistent pathology are major determinants of outcome, with initial primary anastomosis frequently being associated with more complications and a worse outcome.

73.7 Ethical Issues

As in any resource-poor environment, the need for parenteral nutrition may be the major determinant for survival.

73.8 Evidence-Based Surgery

Available literature on colonic atresia is currently in the form of relatively small case series of between 9 and 15 cases and seeks to identify associated conditions, management options, and predictors of outcome. Tables 73.1, 73.2, and 73.3 reflect current available literature.

Table 73.1 Evidence-based literature [13]

Title	Colonic atresia: spectrum of presentation and pitfalls in management. A review of 14 cases
Authors	Cox SG[1], Numanoglu A, Millar AJ, Rode H
Institution	Department of Pediatric Surgery, Red Cross War Memorial Children's Hospital & School of Child and Adolescent Health, University of Cape Town, Klipfontein Road, RONDEBOSCH 7700, South Africa
Reference	Pediatr Surg Int. 2005 Oct; 21(10):813–8. Epub 2005 Oct 21
Problem	Pitfalls in the management of colonic atresia.
Intervention	Fourteen cases of colonic atresia seen over a 38-year period are reviewed with particular reference to clinical presentation and pitfalls in management. Seven had Type I atresia, two had Type II, and five had Type IIIa. Ten had associated gastrointestinal anomalies. Management varied considerably
Comparison/control	Six had primary colonic anastomosis. Two of these developed complications due to unrecognized distal hypoganglionosis, two had associated jejunal atresias resulting in short-bowel syndrome, and two had primary anastomosis protected by proximal ileostomies. Seven had a staged repair with initial defunctioning enterostomy with only one complication, an unfixed mesentery that later resulted in midgut volvulus. The only mortality was a patient in which a jejunal atresia repair leaked as a result of a missed colonic atresia. Operative strategy should depend on the clinical state of the patients, the level of atresia, associated small bowel pathology, and exclusion of distal pathology
Outcome/effect	Primary anastomosis would only rarely be advised with a circumspect approach. Long-term outlook, as in small bowel atresia, is generally excellent

Table 73.2 Evidence-based literature [20]

Title	Colonic atresia and stenosis: our experience
Authors	Mirza B, Iqbal S, Ijaz L
Institution	Department of Pediatric Surgery, The Children's Hospital and the Institute of Child Health Lahore, Pakistan
Reference	J Neonatal Surg. 2012 Jan 1;1(1)
Problem	Colonic atresia and stenosis are rare entities. On average, 1 case per year of colonic atresia is being seen in most of pediatric surgical centers, and to date, less than 10 cases of colonic stenosis have been reported
Intervention	The medical record of patients of colonic atresia and stenosis managed during March 2006 to March 2010 was reviewed

Table 73.2 (continued)

Comparison/control	A total of 15 patients of colonic atresia [11] and stenosis [4] were the study population. Four were ascending colon atresia, two at hepatic flexure and transverse colon each, and one at sigmoid colon. Two patients had multiple colonic atresias. One patient of ascending colon atresia also had pyloric atresia. In colonic stenosis population (two congenital and two secondary to necrotizing enterocolitis), two were transverse colon stenosis and two were sigmoid colon stenosis. The preoperative diagnosis was distal small bowel atresia in 11 patients. Colonic atresias were managed by colocolic anastomosis with covering ileostomy in eight patients. The remaining three patients were managed by exteriorizing both ends of atresia. Colonic stenosis cases were managed by primary colocolic anastomosis in one patient and colocolic anastomosis under covering ileostomy in three patients. Three patients of colonic atresia succumbed postoperatively
Outcome/effect	Colonic atresia and stenosis are rare entities. Associated alimentary tract malformations may result poor prognosis. Colonic atresia can safely be managed by colocolic anastomosis with covering ileostomy

Table 73.3 Evidence-based literature [21]

Title	Intestinal atresias: factors affecting clinical outcomes
Authors	Piper HG, Alesbury J, Waterford SD, Zurakowski D, Jaksic T
Institution	Department of Surgery, Children's Hospital Boston, Harvard Medical School, Boston, MA 02115, USA
Reference	Journal of Pediatric Surgery 2008 Jul;43(7):1244–1248
Problem	Despite improvements in care, intestinal atresias are associated with prolonged hospitalization and occasionally mortality. Although each type of atresia is distinct, it is unclear which factors impact clinical course
Intervention	This study seeks to identify predictors of untoward outcome
Comparison/control (quality of evidence)	Neonates with duodenal, jejunal/ileal, and colonic atresia treated at one institution from 1982 to 2005 were reviewed. A total of 132 infants were evaluated, including 63 with duodenal, 60 with jejunal/ileal, and 9 with colonic atresias. Overall mortality was 7% with associated congenital anomalies identified as an independent risk factor. Infants with associated anomalies were more likely to have low birth weight, which further increased mortality risk. Atresia location did not affect mortality or length of stay; however, it did impact the time to full enteral nutrition with jejunal atresia requiring longer than duodenal
Outcome/effect	Colonic atresia (CA) is one of the rarest causes of neonatal intestinal obstructions, and no large series can be reported. Overall mortality from intestinal atresia is low and is not dependent on the location of obstruction. Infants with birth weight less than 2 kg and associated anomalies are at an increased risk for prolonged hospital stay and mortality. Because of the low incidence of CA, delay in diagnosis and treatment may occur. The mortality is statistically higher when the surgical management is performed after 72 hours of age. However, the prognosis of CA is satisfactory if diagnosis and surgical management could be made promptly and properly

Key Summary Points

1. A very large dilated loop with a fluid level on abdominal X-ray is suspicious.
2. Beware the second atresia in the colon of a patient with a small bowel atresia.
3. Exclude Hirschsprung's disease of the bowel distal to the colon atresia.
4. Avoid primary anastomosis.
5. Note the association with nonrotation of the midgut.

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