KISA VE POŞ ŞEKLINDE KOLON

SHORT AND POUCH COLON

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Özet

İmperfore anüs,kısa ve poş şeklinde kolonu olan 2 yenidoğan ve bir infant olgusu sunuldu. Birlike bulunan diğer anomaliler değerlendirildikten sonra tüm olgular opere edildi. Bir olguda çekostomi 2 olguda ileostomi yapıldı. Bu nadir anomalinin klinik özellikleri ve sonuçları embriyoloji ve tedavisine özel bir önem verilerek tartışıldı.

Anahtar kelimeler: *Anorektal malfarmasyon, Kısa kolon, Poş şeklinde kolon, Kloaka*

Summary

Two newborns and one infant presenting with imperforate anus and short and pouch colon are reported. After evaluating the associated anomalies, all patients underwent surgery in which cecostomy (n:1) and ileostomy (n:2) were performed. The clinical features and outcome of this rare anomaly is discussed with special emphasis on embryology and treatment.

Key words :*Anorectal malformations, Short colon, Pouch colon, Cloaca*

AÜTD 1997, 29:478-480 **MJAU 1997, 29:478-480**

Introduction

The association of short and pouch colon with imperforate anus is a rare anomaly. In this unusual malformation, a varying length of the colon is replaced by a dilated pouch which has a wide high fistulous communication with bladder or vagina. Short and pouch colon is usually seen in male gender (1-5). The absence of haustrations, appendices epiploicae and tenia coli are the major anatomic features of this supralevator anomaly (1). The aim of this report is to present two newborns and one infant with this rare anomaly to discuss the embryology and outcome.

Case reports

Case 1:

A 7-day old boy, who was born with an imperforate anus, was admitted, moribund, with abdominal distention and bilious vomiting. Meconium was excreted from his urethra. A clinical diagnosis of high imperforate anus was made. The initial operation was performed using a transverse right upper incision for transverse colostomy because the anomaly was not preoperatively suspected. During surgery for transverse colostomy, the transverse colon was not found at the expected site. At the time of further exploration, it was discovered that the entire colon was represented by a thin-walled dilated pouch lying in the lower abdomen. There was no cecum or appendix. This colonic pouch was 11 cm by 10 cm and had no tenia coli. Because of the patient's unstable respiratory condition, disconnection of the colovesical fistula was not attempted. An ileostomy was performed through left upper quadrant but the infant died due to septicemia on the second postoperative day.

Case 2:

A 4-day-old boy, born with imperforate anus, was admitted with abdominal distention and vomiting. There was a history of passage of meconium and flatus through the urethra. A clinical diagnosis of high imperforate anus with rectourethral fistula was made. X-ray films of the abdomen taken in the upside down position, revealed signs of acute intestinal obstruction with a large dilated segment, this dilatation was thought to be due to prolonged obstruction of the colon. At operation for transverse colostomy, the transverse colon was not found, on further exploration, the colon appeared as a short and extremely dilated pouch filled with meconium. There was no cecum or appendix. The pouch colon was communicating with the bladder but disconnection of the fistula was not achieved from the transverse right upper incision. A loop ileostomy was performed through the left upper quadrant. The child progressed well postoperatively. Subsequently, this case was lost to follow-up.

Case 3:

A one-year-old girl with imperforate anus was admitted with complaint of passage of stool through the vagina. A clinical diagnosis of high imperforate anus with persistent cloaca was made. At operation

Figure 1. *As Seen On Laparatomy, Short and Pouch Colon With Antimesenteric Vascular Supply From the Superior Mesenteric Artery.*

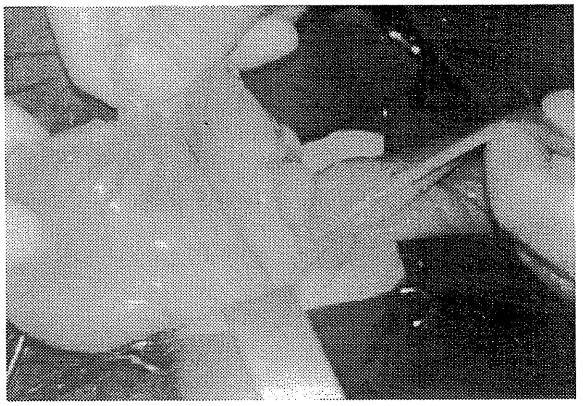
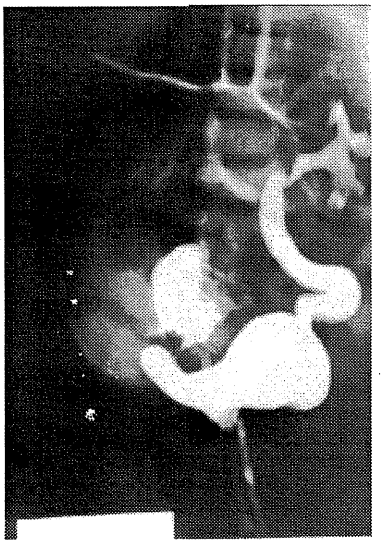


Figure 2. *Cloacogram of Case 3 Showing A Left Vesicoureteric Reflux.*



for transverse loop colostomy, the transverse colon could not be found. Exploration demonstrated intestinal malrotation. The whole colon was represented by a dilated pouch lying in the left lower abdomen above the bladder. There was cecum with appendix. The colonic pouch that communicated with upper and posterior part of the cloaca, was 25 by 16 cm and had no tenia coli. Its mesentery was diminutive, preventing its exteriorisation. (Fig 1). Cecum was exteriorisated through left upper quadrant. A cloacogram was performed at postoperative period.Left vesicoureteric reflux was encountered (Fig 2). The child was under follow-up for subsequent surgical treatment.

Discussion

Imperforate anus is a common disease in children and its association with colonic malformations such as atresia, agenesis and shortening have been described (6-9). Isolated short colon associated with imperforate anus has been regarded as a part of cloacal exstrophy (10). Anorectal agenesis with a short colon in the form at a pouch or sacculer colon is a different entity that appears to be most common in India and China (1,5,11). In this supralevator anomaly, the normal small intestine ends abruptly in an enormous colonic pouch which is 10-15 cm in diameter. Congenital heart disease, sacral agenesis, hypospadias, syndactyly, double or absent appendix, mongolism and vesicoureteric reflux are the most frequent associated anomalies (2,5). A possible etiological factor is non-development of the inferior mesenteric artery. Intra-uterine obliteration of this artery or a part of it probably leads to a stunted and malformed hindgut. The mesentery of the short and pouch colon is short, and there are no haustrations, appendices epiploicae and tenia coli (5). The ileum was seen enter the pouch from right to left and opened at a low position, close to the genitourinary fistula. The other striking anatomical features seen in patients with short and pouch colon are the vascular supply from a terminal branch of the superior mesenteric artery along its left or lateral border (1). In the newborn period, as described in the literature, the transverse diameter of the distended segment exceeds half the transverse diameter of the abdominal cavity at the widest part. This dilatation is explained by accumulation of meconium in this segment starting from the later months of pregnancy (1). In Case 3, transverse diameter of the short colon was not so dilated despite of 1 year period. The fistulous communication was always found to be very wide with easy and free flow contrast from the colonic pouch (4). Explanation of dilatation that is only due to accumulation of meconium seems to be difficult. This characteristic feature was not seen in cases of imperforate anus with normal length of the colon. We often see infants with imperforate anus in rather advanced stages of intestinal obstruction presenting as late as 4 or 6 days after birth providing ample time for the colon to become

distended. For this reason, a pouch colon is an integral part of the short and pouch colon that associates with imperforate anus. This dilatation may be explained by lack of tenia coli in this dilated segment. This anomaly is frequently seen with common cloaca in girls (1).The high incidence of "rectovaginal fistula" reported in the literature actually reflects a high incidence of misdiagnosed cloacas in supralevator anorectal malformations of girl patients (12). For this reason, short and pouch colon which is associated with a supralevator anomaly, is frequently seen with common cloaca in girls. Anyway, this condition is rare in girls in whom although common cloaca is frequently seen, there are very few cases related to this subject in the literature (1,2,4,11). Although the embryology is so confusing, the mode of treatment is a straightforward staged approach. Divertive procedures of the gastrointestinal tract should be followed by the ligation of the fistula and complete removal of the short and pouch colon which is non-functional. Pull-through of the ileum consists the last part of the staged operation.

References

1. Chadha R, Bagga D, Malhotra CJ, et al. The embryology and management of congenital pouch colon associated with anorectal agenesis. J Pediatr Surg 1994;29:439-446
2. Narasimharao KL, Yadav K, Mitra SK, et al. Congenital short colon with imperforate anus. Ann Pediatr Surg 1984;1:159-167
3. Shing S, Pathak IC. Short colon associated with imperforate anus. Surgery 1972;71:781-786
4. Singh A, Singh R, Singh A. Short colon malformation with imperforate anus. Acta Pediatr Scan 1977;66:589-594
5. Wardhan H, Gangopadhyay AN, Singhal GD, et al. Imperforate anus with congenital short colon (pouch colon syndrome). Pediatr Surg Int 1990;5:124-126
6. Blunt A, Rich GE. Congenital absence of colon and rectum. Am J Dis Child 1967; 114:405-406
7. Dickinson SJ. Agenesis of the descending colon with imperforate anus. Correlation with modern concepts of the origin of intestinal atresia. Am J Surg 1967;113:279-281
8. Santulli TV, Schllinger JN, Amoury RA. Malformations of anus and rectum. Surg Clin North Am 1965;45:1253-1271
9. Spencer R. Exstrophia splanchnica (exstrophy of the cloaca). Surgery 1965;57:751-756
10. Magnus RV.Ectopia cloaca-A misnomer. J Pediatr Surg 1969;4:511-519
11. Yuejie W, Rong D, Guie Z, et al. Association of imperforate anus with short colon. a report of eight cases. J Pediatr Surg 1990;25:282-284
12. Pena A. Current management of anorectal anomalies. Surg Clin North Am 1992;72:1353-1416

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