



Congenital Pouch Colon with Anorectal Malformation: A Report of One Case

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Abstract

Congenital Pouch Colon (CPC) is an unusual, rare abnormality in which a pouch-like dilatation of colon might be associated with an anorectal malformation. The pouch has different structural histological and functional characteristics in comparison with a normal colon. It has been classified into various forms based on the length of the involved colon. The curative management might vary from single surgical procedure to three stage surgical correction.

In the current case report, we are going to present a 16-hour old, male neonate with type II CPC accompanied with colovesical fistula.

Keyword: Anorectal malformation; Colovesical Fistula; Congenital pouch colon

Introduction

Congenital pouch colon is an unusual abnormality in which a pouch-like dilatation of a varying degree of shortened colon is associated with an Anorectal Malformation (ARM) [1]. A fistulae formation among the pouch colon and genitourinary tract should be suspected.

CPC is more common in males, with male to female ratio of 4:1. Significant number of patients has been reported from India. The etiology of this anomaly has not been clarified yet. The fistulae tract ending differs among girls and boys. Colovesical fistulae just proximal to bladder neck in male and either Colourethral or Colovestibular fistulae in female patients should be highly suspected [2].

The most widely used classification of CPC was described by Narasimbara et al in 1984. The author has classified the CPC concerning the normal proximal colon [3] (Figure 1).

Tubularization after mobilizing the pouch, (coloplasty), is the surgical procedure of choice. We report herein a neonate affected by type II CPC and colovesical fistula who underwent coloplasty. To the best of our knowledge, similar cases are very rare.

Case Presentation

Patients

A 16-hour old, 3-kg weight, male child with abdominal distension and absence of the anal opening was admitted to our hospital.

He was born by a cesarean section at 37 weeks of gestational age. On physical examination, there was no other congenital anomaly. Abdominal ultrasonography revealed left dilated ureter and plain abdominal X-ray showed massive gas shadow of approximately 90mm in diameter. Based on pelvic x-ray, he had normal sacrum (Figure 2).

Surgical procedure

The patient underwent laparotomy by transverse incision. The intraoperative exploration revealed an obviously distended bowel pouch. The mentioned distended pouch was located distal to normal appearing cecum which was mislocated at left upper quadrant. Obliterated appendix was significantly atrophied (Figure3). The abnormal appearing colon pouch which was characterized by lack of tenia coli terminated to bladder neck. Appendectomy, Ladd's procedure and consequent double barrel ileostomy was undertaken. Thirteen months later, subsequent surgical procedure was performed including fistula closure, stapler coloplasty and pull-through anorectoplasty (Abdominoperineal pull-through coloplasty; APPT). A distal colostogram and plain abdominal

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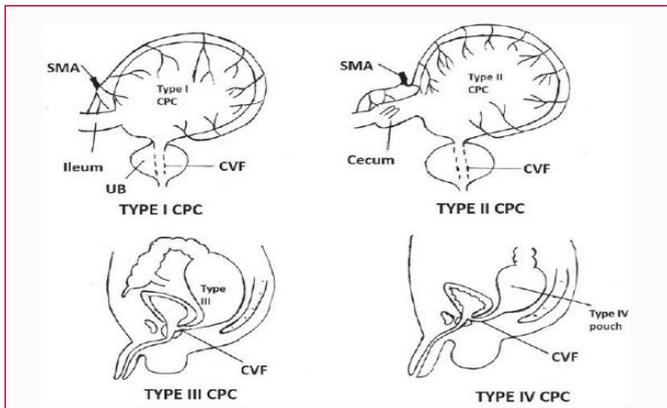


Figure 1: Types of pouch colon based on length.



Figure 4: Distal colostogram followed by a second surgery.



Figure 2: Plain abdominal X-ray showing massive gas shadow about 90 mm in diameter.



Figure 3: Operative view showing a significantly distended pouch-like segment of colon distal to the normal cecum.

X-ray was performed couple of months followed by the second surgery (Figure 4). Ultimately, the ileostomy closure was undertaken as the final step of the three-staged surgical correction.

Discussion

The primary report of a patient suspicious for CPC was done by Spriggs in 1912 from London. He mentioned a patient with the agenesis of colonic distal half combining with rectum, but the CPC nomenclature did not refer to the same time [4]. In 1972, Singh and pathak from India published a case series including six patients associated with imperforate anus and named this anomaly as "short colon" [2]. Narsimha Rao et al., [3]. Suggested "pouch colon syndrome" for this definition and also proposed an anatomical classification of the condition. The incidence of CPC among those affected by high Anorectal Malformation (ARM) varies from 10% to

26%. However, we should keep in mind the 2% to 18% possibility of CPC along with all cases of ARM [5,6]. The significant higher rate of male patients has been well documented in all series [6]. The discrete path physiology and affected phase of fetal life in CPC has yet to be determined.

A large loop of bowel with almost always an air fluid level which has proportionally encompassed more than 50% of the plain upright or inverted X-ray might be diagnostic for CPC. The location of pouch in X-ray might be estimated based the Assumed Pubococcygeal Line (PCL) [6-8]. In the present case, the CPC diagnosis was obtained preoperatively.

The preferred management for CPC has been defined as a multiple surgical procedure including:

- A. Initial operative procedure: proximal colostomy/ileostomy.
- B. Definitive operative procedure: Division of fistula. Coloplasty and Abdominoperineal Pull through (APPT) of coloplasty colon.
- C. Ileostomy/colostomy closure.

In the predicted three-staged procedure, adequate functional lengthening is prepared for absorption and storage [6].

Although one and two-staged procedures are the alternative choices, we planned the three-stage procedure for this patient. Proponents of three stage procedure argue the improved tolerance of these treatment steps during infancy. Accordingly, we performed the final stage after 13 months.

Complication might be related to colostomy, coloplasty, pull through and short length of colon [6].

Complication following coloplasty includes: Suture line leaks, wound dehiscence, dilatation of colon, constipation or watery diarrhea and weight loss the later complication might occur due to the remaining short length of colon [6].

The in hospital post-operative course for 7 days was uneventful. Moreover, the out-patient post-operative surgical clinic at two-week, first and third post-operative month was not significant for possible complications.

Conclusion

Although the incidence of Congenital Pouch Colon is not common, but the possibility of this diagnosis should not be ignored.

The optimal pre-operative and intra-operative diagnosis might change the clinical course significantly as stage procedures might result in better outcomes and less complications. Our patient was referred to a subspecialty Pediatric Surgery center which utilized the intra-operative diagnosis and precise staged management.

References

1. Chadha R, Gupta S, Tanwar US, Mahajan JK. Congenital pouch colon associated with segmental dilatation of the colon. *J Pediatr Surg.* 2001;36(10):1593-5.
2. Singh S, Pathak IC. Short colon associated with imperforate anus. *Surgery.* 1972;71(5):781-6.
3. Narsimha Rao KL, Yadav K, Mitra SK, Pathak IG. Congenital short colon with imperforate anus (pouch colon syndrome). *Ann Pediatr Surg.* 1984;1:59.
4. NJ S. Congenital occlusion of the gastrointestinal tract. *Guys Hosp Rep.* 1912;766:143.
5. Chadha R, Gupta S, Mahajan JK, Bagga D, Kumar A. Congenital pouch colon in females. *Pediatr Surg Int.* 1999;15(5-6):336-42.
6. Devendra K, Gupta SS. Congenital pouch colon-Then and now. *Indian Assoc Pediatr Surg.* 2007;12(1):5-12.
7. Wakhlu A, Wakhlu AK. Technique and long-term results of coloplasty for congenital short colon. *Pediatr Surg Int.* 2009;25(1):47-52.
8. Ghritlaharey RK, Budhwani KS. Two-staged management for all types of congenital pouch colon. *Afr J Paediatr Surg.* 2013;10(1):17-23.