Original Article

Congenital Pouch Colon: A Comparative Study between Two Modalities of Management

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Abstract

Background: Congenital pouch colon (CPC) is an unusual anomaly with an unique geographical distribution. The aim of this study was to find out the incidence of CPC among anorectal malformation (ARM) cases in our institute and to compare the outcome between conventional three-staged surgery versus two-staged management approach. **Materials and Methods:** This study was conducted in the department of pediatric surgery over a period of 7 years from 1st April 2013 to 31st March 2020. **Results:** Out of 754 cases of ARMs, 43 cases of CPC were detected. The incidence of pouch colon among patients with high ARMs was found to be 7.6% with a male predominance (M:F = 4.4:1). The anomaly was diagnosed in 72% of our patients preoperatively and Type IV variety was the most common intra-operative finding. The survival after initial hospitalisation was 82% and 88% in three-stage and two-stage surgical procedures, respectively. However, the final clinical outcome after the completion of all stages of surgery and follow-up was better in two-staged approach (54% vs. 47%). **Conclusion:** Although, CPC is a rare anomaly, the incidence in our institute is 7.6% among high ARM cases. As compared to conventional three-staged surgery, the two-staged management approach has the advantage of better survival and decreased morbidity.

Keywords: Abdomino-perineal pull through, anorectal malformation, colovesical fistula, two-staged management

Introduction

Congenital pouch colon (CPC) is an unusual anomaly where the whole or part of the large bowel exhibits a pouch-like dilation and communicate distally to the urogenital system by means of a fistula. The condition is associated with anorectal malformation (ARM) and is also mentioned as 'congenital short colon' or 'pouch colon syndrome'.[1] CPC is known for its unique geographical distribution in the world. More than 90% of cases are reported from India, especially from the North-West regions.[2,3] Few cases are reported from neighbouring states such as Bangladesh, Nepal and Pakistan and sporadic cases from Japan, China and United Kingdom.[1] Although our hospital is located in the eastern region of India, we came across a good number of cases of CPC. However, a precise study on this congenital malformation was lacking from our part of the country.

The standard management of CPC is a three-staged surgical procedure: stage-I: Initial proximal diversion; colostomy/ileostomy, Stage-II: Definitive pull-through procedure with or without excision of the pouch and Stage-III: Colostomy or ileostomy closure. A two-staged management of CPC was started in some centres and authors published their encouraging results.[4] We have performed the two-staged approach in some of our patients. It consists of: stage-I: Excision of the pouch, closure of urogenital tract fistula and end colostomy/ileostomy, Stage-II: Definitive pull-through procedure and abdomino-perineal pull through (APPT). However, a study was needed comparing both the modalities of management. The aim of the present study is to find out the incidence of CPC among all cases of ARM in our tertiary care institute and to compare the outcome between conventional three-staged surgery versus two-staged management approach.

Materials and Methods

The present study was carried out in Pediatric Surgery Department of SVP PG Institute of Pediatrics, SCB Medical

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College, Cuttack, Odisha, India. The Institutional Ethics Committee has approved study vide IEC No. 33/2020/SCBMC, Cuttack. It was a retrospective, comparative study on patients of CPC from 1st April 2013 to 31st March 2020. All the cases of ARM admitted during these 7 years were reviewed. The incidence of CPC among the cases of ARM and high ARM was studied. The detail history, clinical examination of CPC cases along with imaging studies, surgical procedures and complications were analysed. Plain X-ray abdomen erect and cross-table prone lateral (CTPL) view was the principal diagnostic procedure performed during initial hospitalisation. The classification system described by Narsimha Rao *et al.* was followed.[5]

- Type I: Normal colon is absent and the ileum opens directly into the colonic pouch
- Type II: The ileum opens into a short segment of caecum which then opens into the pouch
- Type III: Presence of a significant length of normal colon between the ileum and the colonic pouch
- Type IV: Presence of near normal colon with only the terminal portion of colon (sigmoid and rectum) converted into a pouch.

Patients were managed by staged operative procedures; conventional three-stage (Group A) or two-staged (Group B). During initial hospitalization, proximal diversion of faecal matter was done, and they were discharged with advice for stoma care. They were assessed during follow-up period with haematological investigations, ultrasonography abdomen, distal cologram, and other investigations according to the need. Definitive operative procedure; APPT was performed after 6 months of age (Stage II) and colostomy/ileostomy closure was done in Group A patients within next 2 months. Outcome was measured in terms of incidence of survival, complications after surgery, continence and parental benefit. A statistical analysis of the results was done using data analysis pack in Microsoft Excel. The two treatment groups were compared using Fisher's exact test, and the results are statistically significant when $P < 0.05$.

Results

During the 7-year study period, 754 cases of ARM were admitted and managed, out of which, 519 cases were male and 235 cases were female; M:F ratio being 2.2:1. Pouch colon was detected in 43 cases, which constitutes 5.7% of cases of ARM and 7.6% of cases of high ARMs. There were 35 male pouch patients and eight female patients; revealing a male predominance of 4.4:1 [Table 1]. The absence of anal opening and abdominal distension were the usual clinical presentation. Preoperative X-ray was suggestive of CPC in 31 cases. Plain radiography showing a dilated loop of intestine with air-fluid level occupying >50% of width of abdomen was taken as criteria for the diagnosis [Figure 1]. In rest of the cases, CPC was the intraoperative finding as laparotomy was undertaken on acute emergency basis without delaying for radiology.

Table 1: Incidence and distribution of congenital pouch colon among anorectal malformation patients

Serial No.	Variables	Anorectal malformation	Congenital pouch colon (%)
1	Total number of cases	754	43 (5.7)
2	High ARM	565	43 (7.6)
3	Males	519	35 (6.7)
4	Females	235	8 (3.4)
5	Male-female ratio	2.2:1	4.4:1

ARM: Anorectal malformation

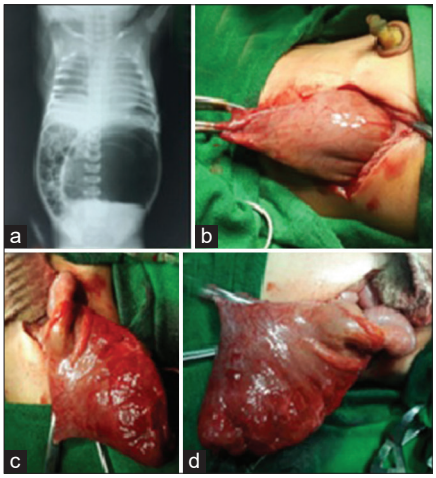

Figure 1: A case of Type I pouch colon (a) Plain X-ray showing large gas shadow on left side of abdomen with air-fluid level occupying more than half of the abdominal width and small bowel loops on the right side, (b) Intra-operative picture of the pouch, (c and d) Ileum opens into the pouch

The intraoperative pathology consistent with CPC was a large thick walled distal bowel with abrupt change in calibre to a pouch communicating to urogenital system by means of a fistula [Figure 2]. Type IV was the most common anomaly found in 31 cases. Type I, Type II and Type III anomalies were found in four, one and six cases, respectively. Among the female infants colovesical fistula was found in three cases and colocloacal fistula was found in two cases. Perforation of the pouch with faecal peritonitis was found in four cases. Associated anomalies were detected in six cases; Down's syndrome, malrotation of gut, horse-shoe kidney, stricture urethra and right side renal agenesis.

Laparotomy and diversion (colostomy or ileostomy) proximal to the pouch were done in 17 cases during initial hospitalization (Group A). Mean age at presentation in this group was 2.52 days, and mean weight was 2.55 kg. Three new-borns died due to septicaemia during hospitalisation. All the 14 discharged cases reported for follow-up initially, but subsequently four cases have not attended [Table 2]. Complications such as colostomy prolapse (two cases) and skin excoriation (three cases) were adequately managed. Parents were advised for colostomy care and distal stoma wash at home. Distal cologram and ultrasonography of the abdomen were done in all cases in the interval period. Finally, eight cases

Table 2: Comparison of outcome between two treatment groups of congenital pouch colon

	Group A (%)	Group B (%)
Initial hospitalization (1st stage surgery)		
Total number of patients	17	26
Survival after first-stage procedure	14 (82)	23 (88)
Complications during follow-up*	5 (29)	6 (23)
Definitive procedure (2nd-stage surgery)		
Patients attended for follow-up (after initial hospitalization)	14	23
Patients undergoing definitive procedure	8 (57)	15 (65)
Survival after definitive procedure	8/8 (100)	14/15 (93)
Survival after 3rd stage surgery (Group A)	8/8 (100)	
Final clinical outcome#	8/17 (47)	14/26 (54)

*P=0.72 (Fishers exact test), #P=0.75 (Fisher's exact test)

have undergone definitive procedure and APPT was done. The mean age at definitive surgery was 1 year and 7 months and mean weight was 9.1 kg. All these cases have undergone colostomy closure within 2 months of pull through procedure. Wound infection, which occurred in one patient after stoma closure was settled. There was no mortality following definitive procedure and following colostomy closure. They have completed 6 months to 2 years of follow-up. Faecal continence was assessed on five patients of this group by Kelly's method and found to be good in three patients and fair in two patients. At present, all patients are asymptomatic, and parents are satisfied regarding the quality of life of their children.

Laparotomy, excision of pouch, ligation of fistula to urogenital tract and end stoma was done in 26 cases [Figure 3] during initial hospitalisation (Group B). Mean age at presentation in this group was 2.73 days, and mean weight was 2.53 kg. There were three deaths, and rest of the cases were discharged with adequate advice. The patients were followed with haematological, biochemical investigations and ultrasonography of the abdomen [Figure 4]. Peristomal skin excoriation noted in four cases was managed and two patients underwent revision of stoma for stenosis. Although all discharged cases attended outpatient department for follow-up in the initial period, five patients did not report thereafter. A total of 15 children have undergone APPT procedure, and all of them were discharged in good condition. Mean age at pull through was 1 year 5 months, and mean weight was 9.5 kg. They were followed from 6 months to 2 years and are doing well. Two cases developed anal stenosis and improved with dilatation. However, one patient was readmitted for septicaemia 3 months after definitive surgery and subsequently died. Continence assessment on eight patients in this group was recorded as good in five patients and fair in three patients. Parents were quite satisfied regarding the growth and quality of life of their children.

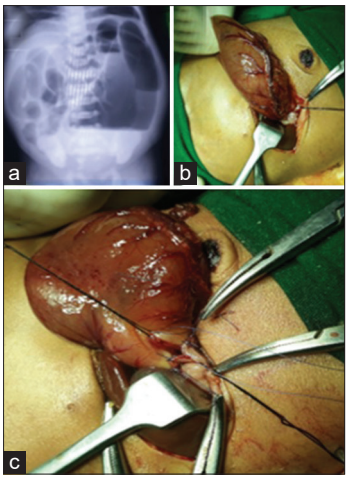

Figure 2: Photograph of a case of type IV pouch colon (a) Plain abdominal radiograph in erect view showing colonic pouch occupying >50% of width of the abdomen, (b) Intra-operative picture of the pouch, (c) Abrupt change of calibre of the colon into a pouch

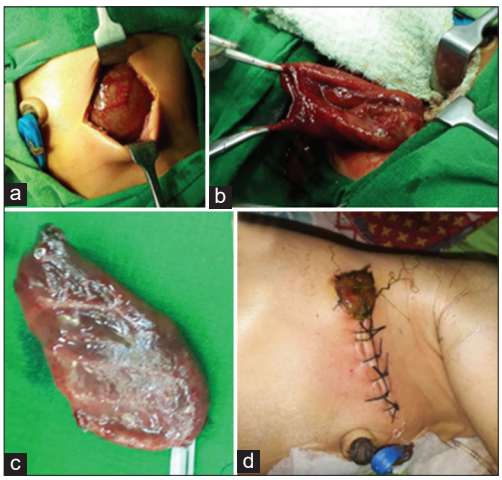

Figure 3: Photographs of a 3-day-old male (group B) undergoing pouch excision and end colostomy: (a) Pouch detected during laparotomy, (b) Pouch mobilized, (c) Excised specimen, (d) End Colostomy

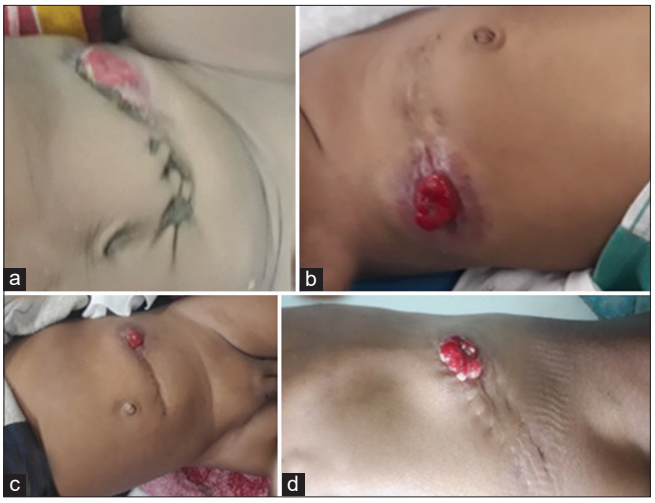

Figure 4: (a) End stoma following excision of pouch, (b) Peristomal skin excoriation, (c) Stenosis of stoma, (d) Functioning colostomy in left lower quadrant of abdomen

Discussion

CPC is associated with a unique geographical distribution in the world. Most of the reports and series are published from India, especially North West regions of the country.[3] However, the malformation was first reported in a specimen of London hospital museum in 1912.[6] The first Indian report was published only after 60 years in a series of six cases by Singh and Pathak, and the anomaly was mentioned as 'short colon'.[7] A precise classification on this malformation was coined by Narsimha Rao *et al.* in 1984. They named this anomaly as 'pouch colon syndrome' and classified into four types.[5] Latter on other classification systems were described and with addition of a Type V anomaly; 'double pouch with a short segment of normal interpositioned colon'.[8,9] However, the classification by Narsimha Rao *et al.* is still widely used and was followed in our centre. Again, Type V anomaly is a rare variety and was not detected in our series. Literature from northern India describe the occurrence of this anomaly among 4.38% to 18.71% of cases of ARMs.[3] The incidence is high in Chandigarh, Delhi, Kashmir, Lucknow, Varanasi and decreases toward the eastern regions of country.[1] Although our hospital does not belong to North West regions of the country, we came across a good number of cases of CPC. To the best of authors' knowledge, this is the first series and study on CPC from our state, Odisha, which belongs to the eastern region of the country and having a population of 45 million. CPC comprised of 5.7% of all cases of ARM and 7.6% of high ARM in our study. This incidence comes within the range of previously reported literatures from the northern parts of the country.[10] Associated anomalies such as malrotation, Down's syndrome, renal agenesis and stricture urethra encountered in our study were similar to other reports.[1]

The malformation most often affects male infants with a male-to-female ratio around 4:1 in different series.[11-13] We have also found a male predominance of 4.4:1 (M:F) in our study. All cases were associated with high ARM, similar to the findings by other studies.[4] However, Pavai *et al*. reported three cases of CPC in association with low ARM, but low ARM in association with CPC was not found in our study.[14] Plain erect abdominal radiograph and traditional invertogram/CTPL pictures are enough to diagnose a case of CPC.[3,13] CPC was detected in 72% of our patients preoperatively. The classical radiological findings are missed in the presence of free gas under diaphragm caused by perforation of the pouch. A new-born with early perforation and high ARM from a region with high prevalence of pouch colon may point toward CPC.[1] Four such cases were encountered in this study. There is a gradual change of morphological pattern of CPC cases. Type I variety was found to be the most common in earlier literatures on pouch colon.[14,15] However, Type IV variety is more frequently reported in recent studies.[12,16-18] Type IV anomaly was detected in 72% of our cases. Our study in combination with the current literatures suggests a changing scenario regarding the most common pathological type of the anomaly and the resultant decrease in severity.

The classical management of CPC is a three-staged surgical procedure[4] where Stage I involves any one of these procedures (i) Colostomy proximal to the pouch, (ii) Window colostomy, where the anterior wall of colonic pouch is opened as stoma, (iii) Division of fistula, coloplasty and stoma, and (iv) Proximal ileostomy; loop or divided. Stage II is the definitive surgical procedure; division of urogenital tract fistula, APPT with or without excision of pouch and Stage III is colostomy/ileostomy closure. Although several operative options are mentioned for initial procedure, the appropriate management should depend on the general condition of the new-born, exact intra-operative pathology and availability of infrastructure. The patient may attend with a colostomy done somewhere else by general surgeons, who are unaware about the anomaly.[4,12] Higher rate of complications is expected in this situation. None of our patients have undergone prior colostomy at other centres. A window colostomy may be a simple surgery with minimal anaesthesia and is required for sick new-borns to decrease the operative time. However, it is associated with unacceptable high mortality (15%–20%) and should be condemned.[1,19] Window colostomy was not performed in our patients. Coloplasty is a procedure where the pouch is tabularised to maintain the colonic peristalsis and absorptive function. Mortality following coloplasty and end colostomy in neonatal period is very high; 30%–50%.[2,5,20] Preserving the colon by coloplasty during definitive pull through procedure may have complications especially without protective stoma.[4] Coloplasty/colorrhaphy was not done in our patients and pouch excision was done.

Although 82% of patients survived initial hospitalisation in Group A, only 57% of these cases have undergone APPT followed by colostomy closure. None of these patients required re-exploration or admitted for post-operative complications. The probable cause of non-attendance for definitive surgery in rest of the cases may be (i) They are waiting with functioning colostomy to report at a later age due to ignorance, family or financial problems, (ii) Succumbed to colostomy related complications or medical illness, (iii) Attending other pediatric surgical centres. The major advantage of the three-staged surgery is that, the definitive surgery is performed with a proximal protective stoma.

The two-staged management was successfully tried and found to be has certain advantage. Although, Ghritlaharey and Budhwani reported two-stage management in 11 cases of CPC over a period of 12 years, our two-staged management is quite different in terms of operative steps. They have performed colostomy without pouch excision in first stage and pouch excision along with APPT in the second stage.[4] However, ligation of urogenital tract fistula, pouch excision and end stoma was done as first-stage surgery in our series. Excision of pouch and end colostomy should be the choice of initial operative procedure.[1] The stoma should be planned in left lower quadrant in this approach. During the definitive procedure, the end stoma was pulled down as APPT. The survival after initial hospitalisation in Group B was 88%, and

65% of these patients have undergone definitive surgery. The reason for non-attendance of eight patients with end stoma may be similar to Group A. We presume that, they might have attended other hospitals or died due to medical illness/colostomy-related complications or will attend us in future. The two-staged procedure avoids another step of stoma closure as the functioning stoma is mobilised and pulled through during definitive surgery. Hence, the stoma-related complications, morbidity during third surgery, hospital stay and associated costs are omitted. Although, definitive surgery is performed without a protective stoma in this procedure, it is well tolerated in infants at an age of few months.

Primary single-stage management in CPC was also practiced by some surgeons, and they found to have good/fair continence.[18,21] However, due to high risk of complications and mortality, single-stage surgical approach is not advised by many authors.[2,4] Again, definitive surgery at few month later is well tolerated and is associated with better result.[13,22-24] Single-stage surgery was not performed in this series, and authors strongly believe in staged approach for CPC. Complications after initial diversion and following definitive surgery occurred in both the groups, which were adequately managed. Statistical comparison of complications between the two groups was done and was not significant ($P = 0.72$). There was no mortality after definitive procedure in both the groups. Although statistically non-significant ($P = 0.75$), the overall incidence of survival was higher in Group B (54% vs. 47%). Group A patients needed another phase of hospitalization after definitive pull through procedure. In other words, the conventional three-stage surgery is associated with higher morbidity and longer duration of hospitalisation for children. All the patients are reporting quite regularly after final-stage operation. Faecal continence assessment was done by Kelly's method in both the groups and was found to be good/fair.[18,25]

There is a gradual decrease in the mortality from 40% to 15% in this anomaly over the years.[1,2] This is due to improved knowledge about the anomaly, neonatal care and refinement of surgical techniques. There was 14% mortality in neonatal period in our study. Pouch perforation followed by peritonitis and septicaemia is particularly associated with increased mortality. Although studies on pouch colon is continuing, some unsolved questions are yet to be answered, (i) Why a particular geographical area of world is unusually affected and (ii) Role of genetics in the embryogenesis and etiopathogenesis of this peculiar malformation. However, a recent study on whole-exome sequencing revealed the association of CPC with rare mutations and variants.[3]

Limitations

The major limitation of this study is low sample size, which is due to rarity of this congenital malformation. Again, it is a one-centered study, having its inherent problems. Further multicentric study on this congenital anomaly is needed, especially focusing on genetic and environmental factors.

Conclusion

CPC cases account for a significant percentage of ARMs in our institution with gross male predominance. Most of the patients can be diagnosed pre-operatively by strong clinical suspicion and simple radiology. There is a trend for changing scenario from more severe forms of the malformation to favourable pathological types. As compared to conventional three-staged operation for CPC, the two-stage management approach has the advantage of better survival, shorter hospital stay and morbidity in these children. Although the first-stage surgery in two-staged approach was somewhat extensive in new-born period and against the concept of 'initial minimal management', it was well-tolerated and associated with good outcome. Parents of both the groups are satisfied regarding the quality of life and growth of their children, but parental satisfaction was better in two-staged approach due to avoidance of another/third stage of hospitalization. The survival of these infants can be further improved by increased awareness about this congenital anomaly, early referral to tertiary care centres and timely surgical intervention.

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Conflicts of interest

There are no conflicts of interest.

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