Original Article

Congenital Pouch Colon: Our Experience with Coloplasty

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Abstract

Background: Congenital pouch colon (CPC) or congenital short colon is an entity found mainly in Indian subcontinent. In CPC, colon is replaced with partially or completely abnormal pouch connected to the genitourinary tract by a fistula (colovesical). Management protocol is different in different institute. In this article, we are sharing our three stage standard management approach; principle and technique of coloplasty in cases in which colon length is not adequate for pull through and their follow-up. This study aims to show the result of coloplasty in complete CPC. **Materials and Methods:** This is retrospective observational study, of 5 years duration. The medical record of these patients was reviewed for demographic information, clinical features, investigations performed, operative notes, post-operative events and the outcome of surgery. **Results:** Total of 626 ARM cases were managed in 5 years duration in which 64 were of pouch colon. The age of presentation was 1–15 days. In fifty patients who completed their, all stage in that 34 patients were in which coloplasty were done in rest of 16 cases excision of CPC and colonic pull through done in view of adequate colonic length (type III and IV) for pull through. In 34 patient in which coloplasty were done showed satisfactory cosmetic and functional out came after stoma closer in follow-up. **Conclusions:** Properly created coloplasty and three stage procedure for complete pouch colon give better result and less complications. Excision of pouch is not requiring in all cases of CPC.

Keywords: Anorectal malformation, coloplasty, congenital pouch colon

Introduction

Congenital pouch colon (CPC) is a well-known variant of anorectal malformation (ARM) endemic in North India.[1] Management of this condition is still evolving. The different institute follows their own management protocol. Management of type III and IV pouch colon is straight forward with excision of pouch and colonic pull through. There is a lot of debate in literature on management of CPC type I and II. Some centre in India and abroad favour excision of pouch colon and ileal/colonic pull through; others favour preserving the part of pouch for coloplasty.[2-7] Both the above approach of managements can be done in one, two or three stage depending on surgeon's experience and institutional protocol. The advantage and disadvantage of both approaches were reported in literature. The major disadvantage of ileal pull through are severe perineal excoriation and frequent stool and in coloplasty are constipation and dilatation of preserved part of CPC.[8] Although some reports suggest better to excise the pouch completely whatever type was because histopathological examination of the pouch revealed abnormal muscle coat with failure to produce propulsive movement but in our experience, it is better to use part of pouch in complete pouch colon for water absorptive and reservoir purpose.[9-11]

Materials and Methods

This is retrospective observational study, of 5-year duration (January 2009–January 2015). Patient admitted to authors unit in paediatric surgery department with CPC were included in this study. The medical record of these patients was reviewed for demographic information, clinical features, investigations performed, operative notes, post-operative events and the outcome of surgery. Follow-up of patient was 6 months to 4½ years after complete procedure was done.

Our principle and technique of coloplasty

Our practice of initial surgery at neonatal period of diversion of stool by colostomy if proximal colon is available, window colostomy in which cases where proximal colon were not

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Access this article online

Quick Response Code:

Website: www.afrjpaedsurg.org

DOI: 10.4103/ajps.AJPS_88_16

How to cite this article: Singh S, Rawat JD, Kumar P. Congenital pouch colon: Our experience with coloplasty. Afr J Paediatr Surg 2018;15:16-21.

available or proximal ileostomy in cases where pouch is perforated. Second stage of surgery were done when baby were fit for safe anaesthesia and able to tolerate major surgery of coloplasty at an average age of 3 months. Abdomen was opened through left lower abdominal incision depending on previous incision. If colonic length was found adequate (6 inches) then pouch was excised and pull through of residual colon done. In cases of inadequate (<6 inches) colonic length [Figure 1] as in type I and II CPC coloplasty was done. Our principle of coloplasty was lumen should be equal to proximal ileum/colon. The length of coloplasty should not be more than 6 inches. In general, type I, II pouch was supplied by superior mesenteric vessels and type III, IV were supplied by inferior mesentery vessels. During mobilisation of pouch, after disconecting window colostomy and fistula [Figure 2], need ligation of inferior mesenteric vessels if present and some branches of superior mesentery vessels so that adequately pull the coloplasty segment up to perineum. Tube should be created on mesenteric side with excision of extra part of pouch on anti-mesenteric side hand sewing or using GIA stapler as shown in Figure 3. Pull through of tabularise colon will be done after creation of space of sacrum followed by posterior sagittal anorectoplasty (PSARP). Patient kept on proximal diversion on colostomy or ileostomy. In third stage of surgery after assessing the distal colon with contrast study [Figure 4], colostomy/ileostomy closer was done. All patients were kept in follow-up. Factors monitors in follow-up are voluntary bowel movement, soiling, continence, constipation, perianal excoriation, features of colitis and growth of children.

Results

Total of 626 ARM cases were managed in 5 years duration in which 64 were of pouch colon. Most of cases were from Uttar Pradesh and Bihar and from low socioeconomic strata, area from agricultural background. The ratio of male and female of pouch colon cases were 7:1 (56/8). Age of presentation was 1–15 days (average - 3.77 days). Presentation of CPC in our study were absence of anal opening in all cases, abdominal distention 65% (38/59) cases, bilious vomiting in 48% (28/59) cases, 45% (23/52) of male patient present with meconurea. There was no family/sibling history of pouch colon cases. Diagnosis was made before initial surgery with plain abdominal radiograph, gas shadow occupying >50% of abdominal width as shown in Figure 5, in 81% (48/59) cases. In our study numbers of CPC cases in type I, II, III and IV as 33, 11, 6 and 14 as shown in Table 1. Five cases (four male and one female) referred from other centre after initial surgery four with colostomy and one with ileostomy. Primary surgery was window colostomy in 38 cases, colostomy in 11 cases and ileostomy in ten cases. Five cases of type I pouch colon presented with perforation peritonitis in these cases ileostomy were done with repair of perforated pouch. In 91% (51/56) of male cases, there were colovesical fistula found and in rest of five cases fibrous band or no fistula found. Out of eight female patients, vestibular fistula found in three patients, and in rest of five cases single perineal opening found in whom on further evaluation three cases had colovesical fistula and two cases had fistula anterior to vagina, posterior to urethra.

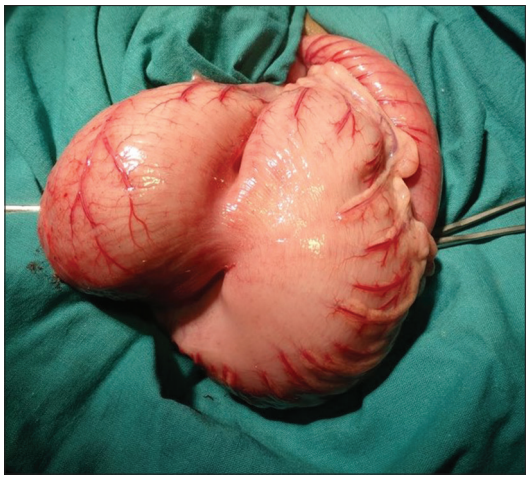

Figure 1: Intraoperative photograph of congenital pouch colon

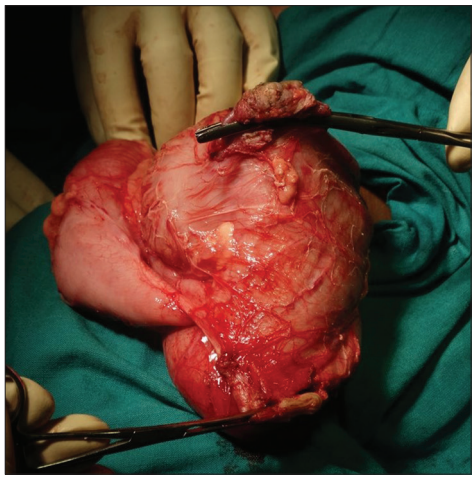

Figure 2: Intraoperative photograph of congenital pouch colon showing disconnected fistula and window colostomy site

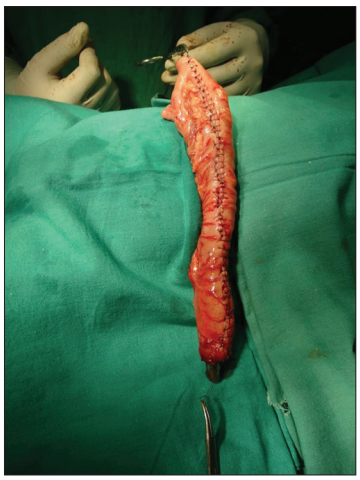

Figure 3: Intraoperative photograph showing coloplasty of congenital pouch colon

Table 1: Showing type of pouch colon, primary, second and third surgery with their follow up

Type of CPC	Number of cases		Primary procedure	Second procedure	Third procedure	Follow up	Functional score							
	Male	Female					Voluntary bowel movement (yes)		Soiling (yes)		Totally continent (yes)		Constipated (yes)	
							No of cases	%	No of cases	%	No of cases	%	No of cases	%
I	26	7	Window colostomy=24, ileostomy=8, ileostomy outside=1	coloplasty done=34, weighting for surgery=6, Expired=4	Completed all stage=28, lost from follow up=6	18 cases type I	11	60%	9	50%	9	50%	2	11%
II	10	1	Window colostomy=9, ileostomy=2			10 cases type II	7	70%	4	40%	5	50%	1	10%
III	6		window colostomy=5, colostomy done outside=1	Excision of pouch and colonic pull through=16, weighting for surgery=4	Completed all stage=14, lost from follow=2	5 cases type III	4	80%	1	20%	5	100%	1	20%
IV	14		Colostomy=11, colostomy done outside=3			9 cases type IV	9	100%	1	11%	9	100%	2	22%
Total	56	8	64			42								

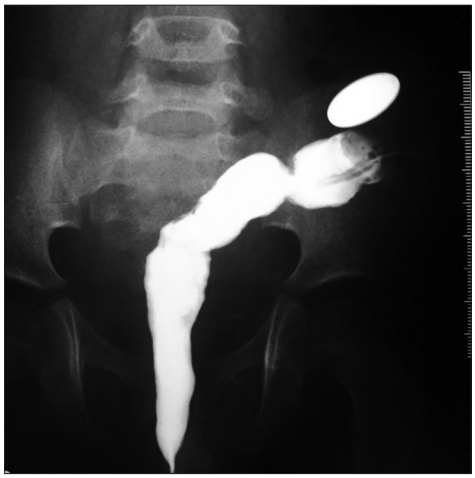

Figure 4: Distal colostograms showing tubularised colon of congenital pouch colon

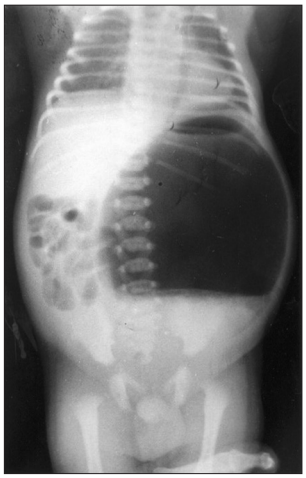

Figure 5: X-ray abdomen of a neonates showing gas shadow occupying >50% of abdominal width

All of female patients had partial or complete vaginal septum with uterus didelphys. In CPC associated anomalies are three hypospadias, one patient has bilateral hydroureteronephrosis, three patients with double appendix, and eight patients with complete CPC had absent appendix, two patients undescended testis as shown in Table 2. Total of fifty patients completed their all stage of surgery, four cases expired after initial surgery due to severe sepsis and associated severe anomalies and remaining (ten) are waiting for completion of stage. In fifty patients who completed their all stage in that 34 patient were in which coloplasty were done in rest of 16 cases excision of CPC and colonic pull through done in view of adequate colonic length (type III and IV) for pull through. Out of 34 of coloplasty group 6 were lost from follow-up and rest 28 were in regular follow-up. Vaginal septum in female patient was excised at the time of pull-through procedure. In our unit, during the study period, we had not done any ileal pull through as our protocol to save the colon.

Discussion

Coloplasty was first described by Trusler in 1959.[12] He had done coloplasty in a neonates with CPC in which 8 cm long colon was created and pull through done with intact vessel. Although the baby expired on day 8 due to sepsis, baby passed stool normally till he survive through neo-anus. Later, the technique of coloplasty describe by Chiba *et al*. in 1976 in two cases successfully with similar technique.[13] In 1982, Vaezzadeh *et al*. had done coloplasty in a female case of CPC with colovesical fistula with excellent result with properly trimmed colon.[14] One year of follow-up, the patient was continent and barium study shows no evidence of colonic dilatation. Subsequently, Chadha *et al*. describe the technique of coloplasty and applied it in type I and II pouch colon in staged surgery of CPC.[7] Initially, he had done this with creating a colonic tube with diameter of 2.5 cm and found dilatation of colonic tube but later he created more

Table 2: Associated anomalies with CPC (*n*=64)

Anomalies	No. of cases
Hydroureteronephrosis/VUR	2
Hypospadias	3
Cryptorchidism	2
Uterus didelphys	8
Partial or complete vaginal septum	8
Double appendix	4
Absent appendix	8
Sacral agenesis	1
Meningomyelocele	2
Meckel's diverticulum	3
TEF + EA	2
Cardiac anomalies	6

narrow colonic tube with a diameter of 1.5 cm with better result. In 1996, the technique of coloplasty was describe by Wakhlu *et al*., he first mobilised the pouch by dividing the inferior mesentery artery, preserving the superior mesentery vessels and creating a tube from pouch on a No. 16 French rubber tube.[6] Excess of pouch tissue were excised. Follow-up of patients were good except colonic dilatation in two cases out of thirty patients. Later, he modified his technique with creation of more narrow tube of colonic strip of 1.5–2.0 cm and preserving inferior mesenteric vessels. Although there is no definite guideline for width of coloplasty, we are doing this with diameter of terminal ileum or proximal colon (in type II) of patients. Some authors have even reported satisfactory results with a patch graft of the pouch colon over the pulled-through ileum in complete pouch colon.[15] We at our centre follow the principle of staged surgery with coloplasty in type I and II CPC to avoid major complications of ileal pull through with satisfactory result on short and long-term follow-up.

The incidence of pouch colon among all cases of ARM is reported between 6.5 and 15.1 in different reports.[2,3,16-19] At our study, the incidence of CPC was 10.2% of total ARM and comprised 23.7% of high ARM. In our study, male patients are 87% and female cases are 12% other study also supports the male predominance.[20] Diagnoses were made before initial surgery in 48/59 (81%) of cases by abdominal radiograph with a big air fluid level occupies more than half of the abdomen. In 11 cases of CPC diagnosis was not made before first surgery as these cases were either type IV CPC or they had very wide urinary fistula. Rest of five cases came from others centre after initial surgery. The type of CPC in our study were type I - 33 (51%), type II - 11 (17%), type III - 6 (1%) and type IV - 14 (21%). The most frequent types of CPC found in other study like Chaddha *et al*., Gangopadhyaya et, and Puri *et al*. were type II and in Tanwani *et al*. study were type I and III.[5,18,21,22]

Single-stage surgery for CPC in neonatal period was reported by Gangopadhyay *et al*. They had done exploratory laparotomy fistula ligation and coloplasty in complete pouch colon with pull through (abdominoperineal pull-through or abdomino – PSARP). As per their reports, advantages of a single stage, procedure includes eliminates urinary tract contamination, establishes anorectal continuity and maximizes the potential for normal defecation reflexes at birth, better continence and cosmesis. Although single stage procedure requires expertise and an 11% mortality due to long duration surgery and high post-operative problems has been reported.[18]

Two-staged procedures were reported by Ghritlaharey *et al*. The initial operative procedure is faecal diversion 2–3 cm proximal to pouch and the definitive procedure includes coloplasty and abdominoperineal pull through without protective stoma. Advantages of this procedure are it provides sufficient time for proper investigations and selection of the cases for two-staged, can be performed for all types of CPC, definitive procedures are well tolerated at few months of age, complications observed following definitive procedures are easily manageable. Disadvantages are definitive procedures are performed without protective stoma may not be a good option when performing a coloplasty as there is always a risk of suture line leak.[23]

Three stage procedures are the standard procedure for management of CPC cases. The aim of surgery is faecal diversion at neonatal period in form of window colostomy/colostomy or ileostomy and coloplasty (in type I, II) with PSARP at second surgery when baby can tolerate major surgical procedure and final stage of stoma closer after healing of coloplasty tube. Reports showed good results in expert hands. Although the technique of coloplasty are different in reports.[2,3,15,17-19,24-26]

In literature technique of coloplasty followed by different authors are different. They found a major problem with coloplasty was colonic dilatation in Chadha *et al*. study 3/9 cases develops this problem, as the technique of coloplasty refined and problem of colonic dilatation lessens in Wakhlu study.[7,19] A recent study published by Sharma and Gupta with long-term 5 years follow-up showed dilatation in 5/7 cases. How the colons were tabularised was not mentioned in their study.[8]

In our study with three-stage procedures, coloplasty had done in total of 34 cases with type I and II CPC. Totally, 28 cases were in follow-up show no colonic dilatation and fair Krickenbeck scoring as shown in Table 1.[27] It is important to exclude anal stenosis in re-dilated coloplasty segment.

Complications of window colostomy in our study were stoma stenosis 18% (7/38), stoma hernia 10% (4/38) and pouchitis 10% (6/59) as shown in Table 3. Patient of stoma stenosis managed with dilatation and washes of stoma, cases of window colostomy prolapse managed with glycerine mops and local care. Cases of pouchitis managed with admission, intravenous antibiotics, and washes of pouch. Complications

following coloplasty are anal stenosis in 10% (3/28), and mucosal prolapse in 14% (4/28) cases. Cases of anal stenosis managed with anal dilatation and cases of mucosal prolapse were managed with mucosectomy during final third stage of surgery of stoma closer. Complications following definitive final third stage of surgery of stoma closer are shown in Table 4. The entire patients in our study gaining weight well, no one had colonic dilatations, and severe perineal excoriations, frequency of stool are three to four times daily. One patient had incontinence due to associated sacral agenesis.

In our experience, cases with CPC in which initial surgery were ileostomy or colostomy instead of window, colostomy had less stoma complication as compare to window colostomy. Moreover, at the stage of coloplasty, it is easy to do in cases where pouch was untouched. Hence, our recommendation for complete pouch colon is to do ileostomy in place of window colostomy at initial surgery.

In our study, mortality after initial surgery in post-operative period was 6% (4/59) due to severe sepsis and associated cardiac disease. No mortality occurs during the second or third stage of surgery.

As in our experience, properly created coloplasty (as described above) and three stage procedure for complete pouch colon give better result and less complications and avoid some severe complications of pouch excision and ileal pull through in which perineal excoriation and malnutrition occurs.

Conclusions

CPC is not rare anomalies in North India. Common presentation is high ARM with abdominal distention just after birth. A supine plane radiograph requires in all cases of high ARM in endemic area to diagnose the condition. Management of pouch colon requires experience and expertise. Excision of pouch is not requiring in complete pouch colon. Proper created coloplasty and placing it in the centre of muscle complex by PSARP route give better short- and long-term result.

Table 3: Complications after initial surgery (*n*=59)

Complications	No. of cases
Wound infection	9
Burst abdomen	2
Window colostomy prolapse	4
Window colostomy stenosis	7
Mortality	4
Pouchitis	6
Recurrent UTI	5

Table 4: Complications after completion of surgery (coloplasty patients *n*=28)

Complications	No. of cases
Episodes of colitis	4
Mucosal prolapse	4
Anal stenosis	3
Colonic dilatation	0
Perianal excoriation	0

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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