Microbiology Section

# A Single Stage Procedure For Congenital Pouch Colon And Its Complications

## A case report

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## ABSTRACT

This is a report of a case of congenital pouch colon (CPC) type II, who was managed by a single stage procedure (exploratory laparotomy, ligation of the colo-vesical fistula, excision of the pouch and abdomino-perineal pull-through of the colon) which was done on the 2nd day of life. He developed severe stricture of the pulled colon and needed three further major operations (exploration and ileostomy, pull-through of the ileum and ileostomy closure). At present, he is 4 years old, is continent, is passing 3-4 stools per day and is doing well.


**Key Words:** Congenital pouch colon, congenital short colon, single stage procedure, surgical complications

## INTRODUCTION

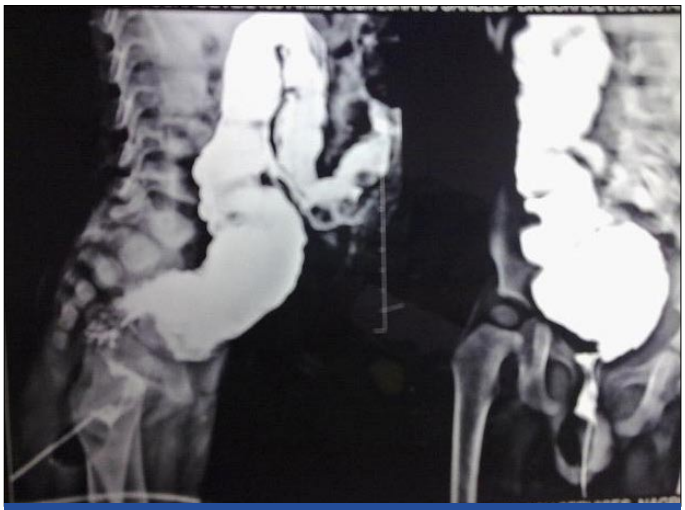
CPC is an anomaly which is associated with anorectal malformation (ARM), where a part or the whole of the colon is replaced by a dilated pouch which communicates distally with a fistula to the genitourinary tract. It is also referred as "congenital short colon", and "pouch colon syndrome" [1], [2], [3], [4], [5], [6]. The standard procedure for the management of CPC is a three stage procedure [1], [2], [4]. A primary single stage procedure for the management of all types of CPC in newborns has also been advocated with better continence, along with low morbidity and mortality [3]. We report herein, a case of CPC type II, which was managed by a primary single stage procedure, but developed severe stricture of the pulled colon and needed three further major operations.

## CASE REPORT

A full term, 3.2 kg, male child was admitted to a nursing home for high ARM with a colo-vesical fistula. Exploration which was done through a left pelvic incision for colostomy; revealed CPC (type II) with colo-vesical fistula and a decision for a single stage procedure was made (not by the authors). Ligation of the fistula, excision of the pouch and the abdomino-perineal pull-through of the colon was done without protective stoma on Oct 3rd, 2006. Postoperatively, he developed perineal wound dehiscence, was treated conservatively and was discharged on the 15th post-operative day. He developed recurrent abdominal distention and features of distal intestinal obstruction due to the stricture of the pulled colon. He was being managed by anal dilatation, enema, saline wash outs, etc, till he presented to us. He also underwent circumcision for phimosis on March 4th, 2008 at the same nursing home by the same surgeon (not by the authors).

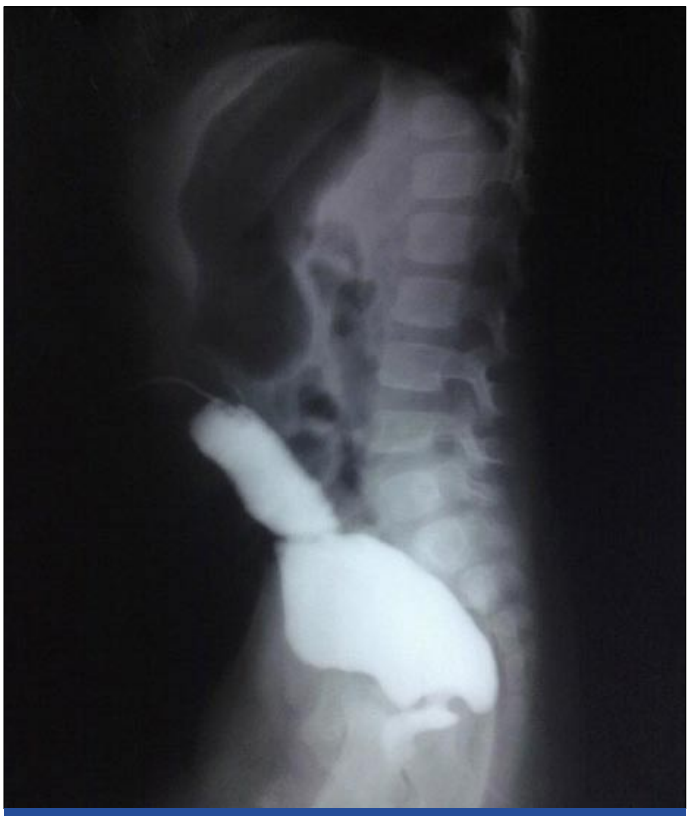
He was referred to us at the age of 21 months with features of intestinal obstruction and a poor urinary stream. His abdomen was distended and visible loops of the bowel were also seen. The anal opening only permitted an infant feeding tube with difficulty and a genital examination revealed meatal stenosis. He already had a contrast enema done, which showed severe stricture of the pulled colon [Table / Fig 1].

The dye study also showed caecum and a dilated distal ileum. On July 17th, 2008, cystoscopy was done and it was found to be normal. He had only meatal stenosis, which was dilated adequately. At the same time, through the right supra - umbilical incision, an abdominal exploration was also done. There were adhesions and dilated loops of small bowel with no normal colon between the caecum and the stricture segment and therefore, adhesiolysis with a loop ileostomy was done. The post - operative period was uneventful. During follow up, he developed stoma stenosis and needed revision after a month. The dye study was done through ileostomy before a re pull-through, which also showed stricture of the pulled colon. The dye study also showed the caecum and the ileum distal to the stoma [Table / Fig 2]. On March 18th, 2010 through an old left pelvic scar, the abdomen was explored and the stricture segment was resected out. The caecum, appendix and the terminal ileum were dissected and mobilized [Table / Fig 3]. As there was no colon for coloplasty and the caecum was also not healthy, we decided to do an ileal pull- through. The caecum with appendix was excised and an ileal pull-through was done. The post operative recovery was uneventful. Four months later; the dye study was done through the ileostomy and it was found to be normal.

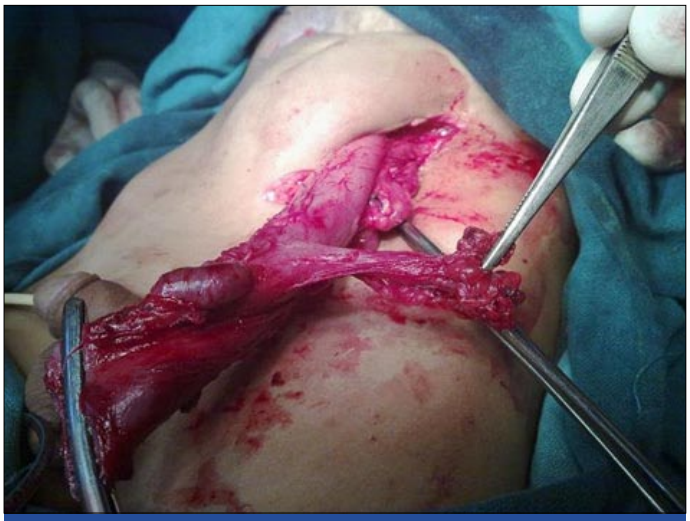


**[Table/Fig 1]:** Contrast enema (AP and Lateral view) showing stricture of the pulled colon. Caecum and distal ileum also seen

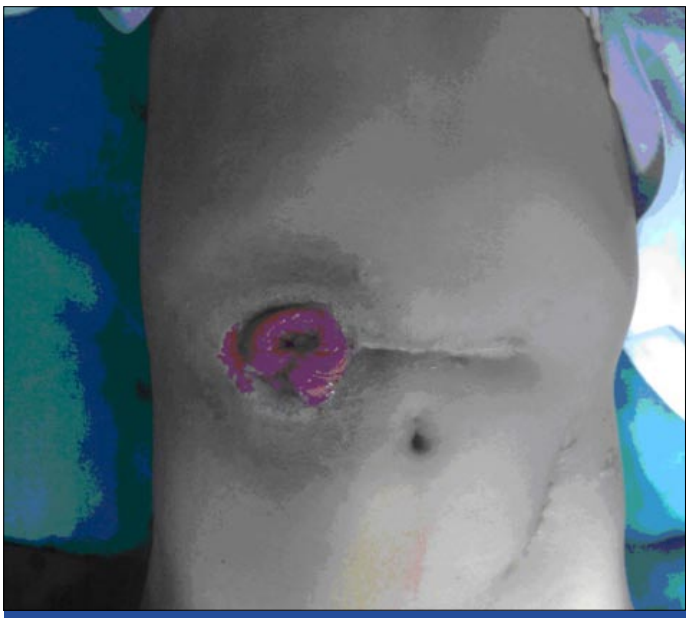
Ileostomy closure was done a month later, on Aug 12th, 2010 [Table / Fig 4] and [Table / Fig 5]. At present, he is 4 years old and is doing well. He is continent and is passing 3-4 stools per day without soiling and perineal excoriation and is in regular follow up.



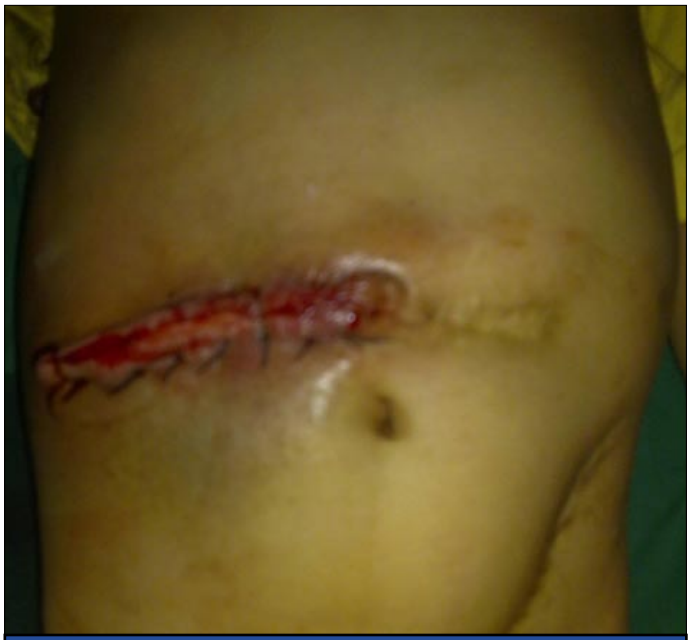
**[Table/Fig 2]:** Contrast study done through ileostomy showing stricture of pulled colon. Caecum and ileum distal to stoma also seen



**[Table/Fig 3]:** Operative photograph showing terminal ileum, caecum and appendix during ileal pull through



**[Table/Fig 4]:** Clinical photograph of patient showing ileostomy and left pelvic scar also seen



**[Table/Fig 5]:** Clinical photograph after ileostomy closure and left pelvic scar also seen

## DISCUSSION

CPC is defined as an anomaly in which the whole or a part of the colon is replaced by a dilated pouch which distally communicates with the fistula to the urogenital tract [1], [2], [3], [4], [5], [6]. Singh and Pathak (1972) called this anomaly as "short colon". Narsimha Rao, et al (1984) named this anomaly as "pouch colon syndrome" and they also proposed an anatomical classification for the same which has been widely accepted [1], [2], [3], [5]. The incidence of the CPC among all the cases of ARM varies from 5 to 17.20% **of all the cases** [1], [2], [3], [6]. The incidence of the CPC among the high ARM cases varies from 10 to 26% **of all the cases** [1], [3]. At our department, we also reported 11.29% of CPC with all ARM cases and 19.60% with high ARM cases [2]. A high incidence (55.8%) of CPC with high ARM cases was reported from Udaipur, Rajsthan [6]. Almost all the series reported male preponderance [1], [2], [3], [5]. This anomaly is seen most frequently in the Northern, North Western, and Central parts of the India and sporadic case reports have also been reported from other parts of the India and the world [1], [2], [3], [4], [5], [6]. The exact cause of the unique geographical distribution is yet to be ascertained.

A widely accepted classification, based on the length of the normal colon, was present proximal to the dilated pouch, as given by Narsimha Rao, et al in 1984 [1]. Wakhlu, et al simplified the classification as "partial short colon" (type A) and "complete short colon" (type B), depending on the length of the normal colon and the need for coloplasty [1], [4]. Recently, Saxena and Mathur (2008) provided a classification for CPC, based on the anatomical morphology and they proposed 5 types of the CPC. They added the type V for double pouch colon with an interposed normal colon segment [6]. The presence of a large air fluid level occupying more than half of the total width of the abdomen on the plain abdominal X-ray, either erect or inverted, is almost diagnostic of CPC [1], [2], [4]. Pre-operatively, it is possible to diagnose pouch colon in about 75 to 90% of the cases [2], [3], [4].

The standard management of the cases of CPC is by staged surgical procedures. The aim of the surgery is to utilize the available length of the colon for absorption and storage capacity, as well as for the capability for propelling the faecal matter onwards with a continent anal opening. In cases with incomplete pouch colon (type III and IV), an adequate length of normal colon is present and so, the pouch can be excised while the colonic function is still preserved (1], [2], [5], [7]. In cases with complete pouch colon (type I and II), these objectives can be achieved only by tabularizing the pouch in the form of Coloplasty [1], [4], [7].

Gangopadhyay et al advocated a primary single stage surgery for all types of CPC in newborns with better continence and cosmesis, along with low morbidity and mortality at low costs [3]. According to them, the single stage procedure includes; exploratory laparotomy and ligation of the fistula, followed by the primary abdominoperineal pull-through or abdomino– posterior sagital anorectoplasty after pouch excision (partial or total pouch colon) or tuboplasty (total pouch colon) [3]. A primary single stage operation at birth relieves alimentary tract obstruction, eliminates urinary tract contamination, establishes anorectal continuity and maximizes the potential for normal defecation reflexes at birth; all these can be achieved at one, rather than multiple operations. To achieve excellent results with the single stage operation, technical expertise is a must. Although a 11% mortality has been reported after a single stage operation, a majority of the mortalities have been attributed to associated malformations [3]. In the present case, the reason for the failure of the single stage procedure; as we believe is the lack of expertise and lack of experience in dealing with cases of CPC. We have been able to achieve good and acceptable results after three stage corrective procedures by using an ileal pull- through.

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