Review Article

Congenital Pouch Colon

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Abstract

Congenital pouch colon (CPC) is an unusual abnormality in which a pouch-like dilatation of a shortened colon is associated with an anorectal malformation. It is categorized into four subtypes (Types I–IV) based on the length of normal colon proximal to the colonic pouch. In males, the pouch usually terminates in a colovesical fistula just proximal to the bladder neck. In girls, the terminal fistula opens either into the urethra or in the vestibule, close to the urethral opening. Girls usually have a double vagina with a wide inter-vaginal bridge, a monocornuate uterus on each side, and urinary incontinence due to a widely open bladder neck. Associated major malformations are uncommon with CPC but sometimes, especially in reports from outside India, major abnormalities are present suggesting an early, severe error in embryogenesis. The more severe Types I/II CPC can usually be diagnosed by a large gas shadow or air-fluid level on X-Ray abdomen. For all subtypes of CPC, it is preferable to preserve a segment of the pouch by fashioning a narrow colonic tube for pull-through, the technique known as coloplasty or tubular colorraphy. Girls need additional management of the genitourinary abnormalities. Postoperatively, fecal continence levels are usually poor, especially with Types I/II CPC.

Keywords: *Anorectal malformation, coloplasty, congenital pouch colon, urinary incontinence*

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Introduction

Congenital pouch colon (CPC)[1] is an unusual abnormality in which a pouch-like dilatation of a varying degree of shortened colon is associated with an anorectal malformation (ARM). The pouch usually terminates in a fistulous communication with the genitourinary tract. The M:F ratio ranges from 2.25:1[2] to 7:1.[3] CPC has been included, as a rare variant, in the Krickenbeck classification of ARM. CPC is much more common in India than in other countries with a review showing that 92.2% of reported cases were from India.[4]

Anatomical Types and Classification

The most widely used classification of CPC was described by Narasimharao *et al*.[5] in 1984 and is based on the length of normal colon proximal to the colonic pouch [Figure 1]. Two other classifications, one by Wakhlu *et al*.[6] based on the length of normal colon proximal to the pouch being < or >8 cm and the other by Gupta and Sharma[3] based on the need for coloplasty, have been described. In essence, "partial" short colon[6] or "incomplete" CPC[3] corresponds with Types III and IV CPC,[5] and "complete" short colon or "complete" CPC with Types I and II CPC.

In 2008, Saxena and Mathur[7] added Type V CPC to Narasimharao *et al.*'s[5] classification where 2 colonic pouches have a short, normal interpositioned colonic segment, the distal pouch ending in a fistula with the genitourinary tract. However, the IAPS classification of 2007[8] in which Type V CPC includes all rarer varieties such as CPC associated with prune-belly syndrome, double colonic pouch, rectal atresia, pseudoexstrophy, and other associations appears more appropriate.

Some recent studies[9-12] suggest a trend of reducing severity of CPC.[10] This has however, not been our experience.[2,4] Type IV CPC also appears more common in reports from Saudi Arabia and Turkey. It is also the

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opinion of Rao and Menon[13] that the incidence of CPC in the West is much higher than the one quoted, as many cases of Type IV CPC are being inappropriately labeled as rectal ectasia/megarectum-megasigmoid.

Anomalous Anatomy

The colonic pouch, especially in newborns, is thin-walled and enormously distended with meconium and flatus.[2,4,5,14] However, with a wide fistula or in older children, it may be smaller and thick-walled. In Type I/II CPC, the ileum or proximal colon usually enters the pouch from right to left in a low position, close to the fistula [Figure 2].

The pouch lacks haustrations and appendices epiploacae. The tenia coli are poorly developed but can usually be made out parallel to the outer margin of the Type I–III colonic pouch [Figure 2]. There is an abrupt transition to normal proximal bowel. In Type I CPC, the appendix is usually absent or else is a small stub. Absence, appendiceal duplication, or a short stubby appendix are frequent with Type II and sometimes Type III CPC.

In Type I/II CPC, the colonic pouch is supplied by a lengthened, hypertrophied terminal branch of the superior mesenteric artery along its outer, lateral aspect [Figure 2]. The inferior mesenteric artery may be absent[1,5,15] but if present, supplies the lower, left side of the pouch.[16] Both mesenteric arteries are present in Type III/IV CPC.

The level of termination of the colonic pouch varies between male and female patients.

Males

The terminal colovesical fistula (CVF) is thought to enter the urinary bladder (UB) midway between its fundus and base,[2,4,16,17] its posterior wall,[5,15,18,19] or at any of three different sites on the posterior wall of the bladder.[3] However, these assertions are not based on conclusive radiological or endoscopic evidence. A recent, as yet unreported, study of 14 boys with Types I–III CPC from our center showed that in all patients, on cystourethroscopic examination, the verumontanum had ascended into the bladder, ending in the distal trigone just proximal to the bladder neck [Figure 3]. In eight patients in whom the CVF had not been ligated earlier, it was seen opening just above and usually to the right of the verumontanum [Figure 3].

In males with Type IV CPC, several reports suggest that the colonic pouch ends in a CVF.[1,3,5,18,19] In another study, we found that in five patients with Type IV CPC, a distal sigmoid colostogram demonstrated a CVF opening just above the bladder neck [Figure 4]. Cystourethroscopy showed the position of the verumontanum and the CVF to be similar to that in Types I–III CPC. The two studies together also showed a large lobulated UB in 7/19 patients (36.8%), hypospadias (n = 5; 26.3%), and undescended testis in three patients (15.8%).

We suggest that Type IV CPC with a CVF just above the bladder neck should be designated as "True" Type IV CPC and differentiated from Type IV CPC associated with an intermediate/low anomaly [Figure 5][2,6,12,14,20] which appears similar to megasigmoid-megarectum associated with repaired intermediate/low ARM.[21,22]

Females

As in males, the subtypes I and II are more common. In a majority of cases, the colonic pouch has been reported to open into either the vagina or in a persistent cloaca.[1,3,5,6,23-26] In other reports, the pouch ended in a vestibular fistula,[6,14,23,26] a CVF,[2,6,14,23-25] a perineal fistula, or a colouterine fistula.[11,23]

In a recent study from our center, the anomalous anatomy in 22 girls with Types I–III CPC was studied

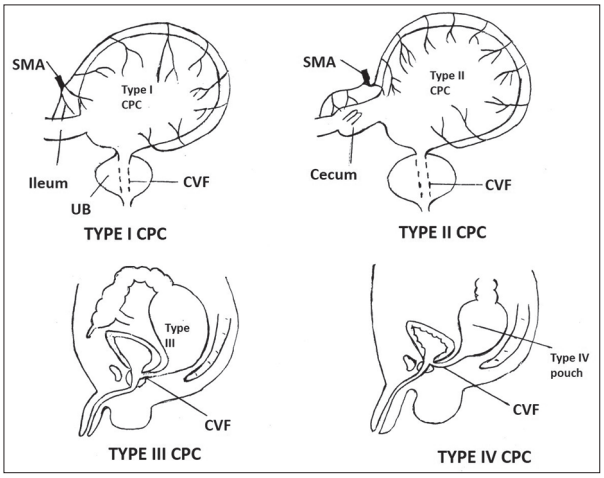

Figure 1: Subtypes of congenital pouch colon.[5] SMA: Superior mesenteric artery, UB: Urinary bladder, CVF: Colovesical fistula

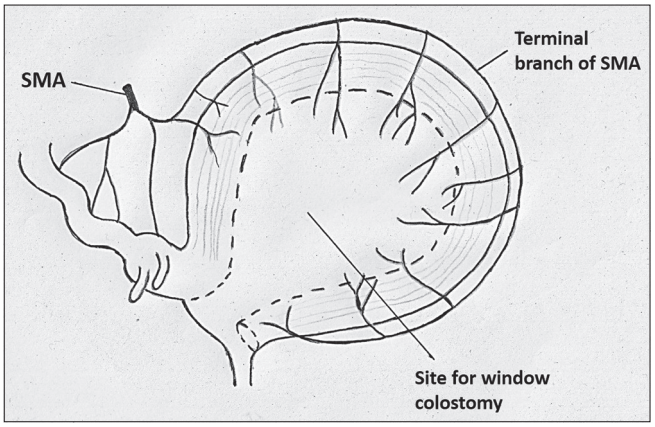

Figure 2: Gross anatomy of Type II congenital pouch colon. Stippled line marks incision for tubular colorraphy. SMA: Superior mesenteric artery

in detail.[27] In contrast to "persistent cloaca," the external genitalia appeared essentially normal with a wide posterior margin of the vestibule [Figure 6]. In newborns, there was often an appearance of folds radiating inwards and upwards from the margins of the vestibule, a "clover-leaf" appearance[24] [Figure 6]. Examination under anesthesia (EUA) showed the confluence of urethral and vaginal openings to be either "low," or relatively "high" with a hypospadic urethral meatus, and double vaginas, usually tilted with a wide inter-vaginal bridge [Figure 7].[27] On EUA and endoscopy, the terminal fistula opened either in the urethra distal to the bladder neck; high in the vestibule, almost "distal urethral" [Figure 7]; or in the vestibule, closer to the perineum.[27] There was usually a short wide urethra, deficient bladder neck, small bladder, poorly developed trigone, and laterally placed ureteric orifices. These features were probably responsible for the high incidence of urinary incontinence (around 85%). All patients had a double uterus with one monocornuate uterus and vagina flanking the fistula on each side.[27]

Apart from a few exceptions,[24,25,28,29] most other studies have not consistently described these findings. This may be because the terminal fistula has a long course beyond the peritoneal reflexion and the gross appearance may erroneously suggest that it is opening into a cloaca, the bladder, uterus, or the vagina.[25,28]

Cases of Type IV CPC with a vestibular fistula and without the features described above have also been reported,[4,12,20] and these appear similar to cases of Type IV CPC with a "low" termination in males.

Histology of the Colonic Pouch

This was thought to be normal in most early reports. In Rao *et al*.'s series,[5] ectopic pancreatic tissue was identified in one specimen. More recently, Agarwal *et al*.[30] reported mucosal inflammation, heterotopic

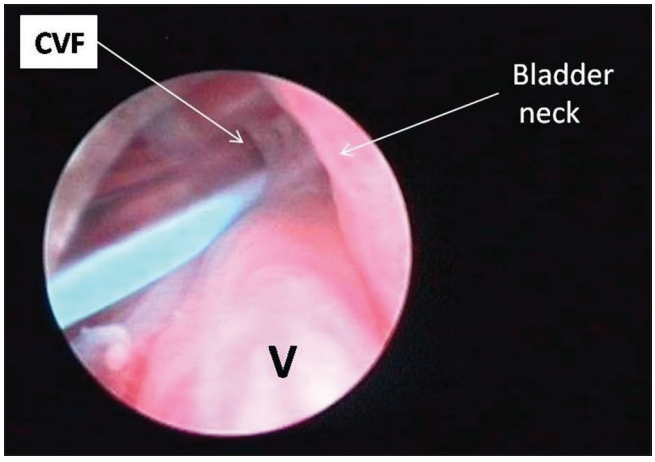

Figure 3: Cystourethroscopy in male with Type II congenital pouch colon showing verumontanum (V) entering bladder neck and the colovesical fistula

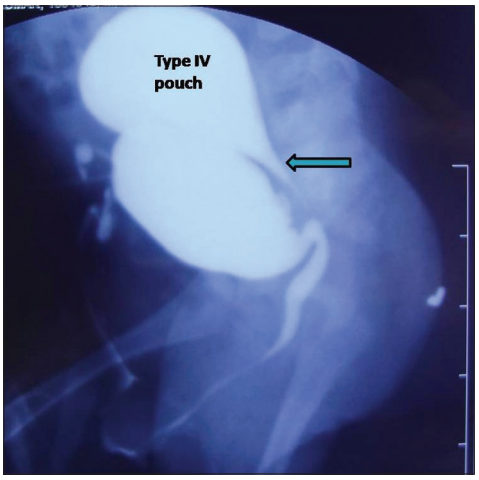

Figure 4: Distal colostgram of a boy with "True" Type IV congenital pouch colon. Arrow points to the long colovesical fistula

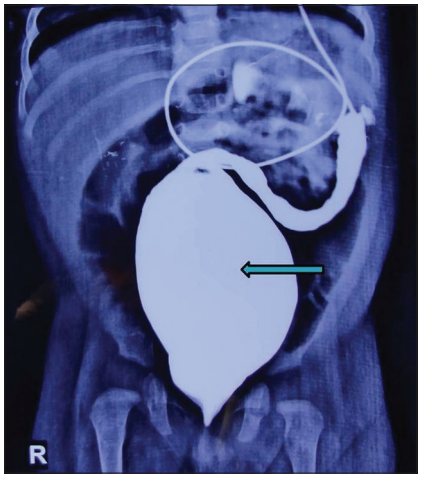

Figure 5: Distal colostogram in a boy with Type IV congenital pouch colon (arrow) and a low termination of colonic pouch

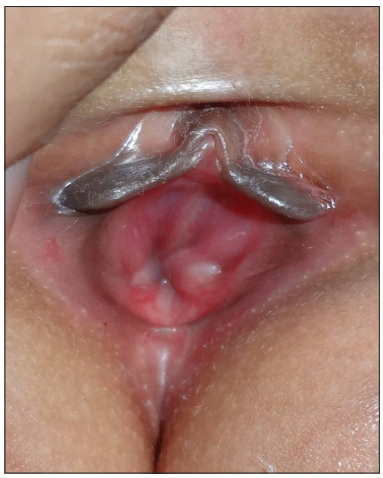

Figure 6: External genitalia in a girl with Type II congenital pouch colon. Note wide vestibule with folds radiating inwards

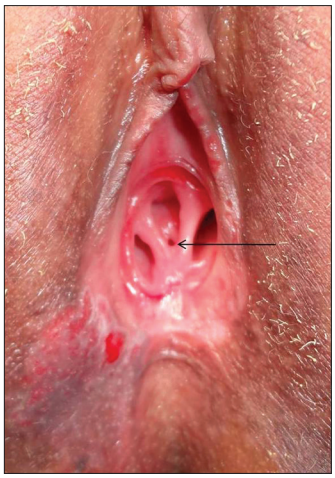

Figure 7: External genitalia in a 10-year-old girl with Type II congenital pouch colon. Arrow shows high vestibular/distal urethral fistula. Note wide urinary meatus, wide inter-vaginal bridge, and excoriations due to urinary incontinence

epithelium (gastric/pancreatic/small intestinal), submucosal fibrosis, thin muscularis mucosae, decussation and splaying of muscle fibers, absent or abnormal (giant) ganglion cells, and neuronal hyperplasia in plexuses. Gangopadhyay *et al*.[31] found fibrosis in all muscle layers on Masson trichrome staining. Chatterjee *et al*.[32] reported a "focal" and discontinuous additional smooth muscle layer. Tyagi *et al*.,[33] in an *in vitro* study on colonic strips from the pouch, found that the tissue failed to show spontaneous contractions but responded to acetylcholine and histamine.

Associated Malformations

In spite of the high supralevator nature of CPC, major associated malformations are relatively uncommon. A review of 566 cases showed hydronephrosis in 40 patients (7%), hydroureteronephrosis (n = 16; 28.2%), vesicoureteric reflux (VUR) (n = 36; 6.36%), and renal agenesis in only 8 patients (1.4%).[11] Other anomalies included esophageal atresia (n = 7) and congenital heart disease (n = 19; 3.4%).[11] Sacral agenesis, usually partial, or other vertebral anomalies were uncommon (n = 24; 4.24%). In CPC, the perineum and sphincteric musculature are also usually well developed.[2,4,6,11,34] A similar low incidence of associated abnormalities has been reported from Pakistan.[18] Our own experience, and other reports suggest a fairly high incidence of hypospadias and undescended testes, and anomalies related to the postarterial limb of the midgut and hindgut such as a Meckel's diverticulum,[4,11] malrotation,[4,11] duplication cyst of the ileum[5,35] or colon,[17] duplication of the colon,[6] and a double Meckel's diverticulum.[14,36]

However, reports of CPC from countries outside the Indian subcontinent,[26,37-39] and occasionally from India,[12,40] show a higher incidence of severe malformations involving both the caudal developmental field and other major-organ systems.

Embryopathogenesis

Smith and Stephens[41] studied male ARM with rectovesical fistula and reported that the verumontanum, vasa and prostatic tissue actually ascend to lie within the bladder at the bladder neck. The findings on cystourethroscopy in males with CPC where the CVF was just cranial to the verumontanum, suggest an even earlier error in the differentiation and growth of the mesenchyme of the urorectal septum (URS), the adjacent developing hindgut, and the postarterial limb of the midgut. The large and flaccid bladder seen in 7/19 boys reported here, as well as in other reports,[6,42] may result from additional involvement of the mesenchyme of the adjacent developing urinary tract.[43] Significantly, at this very early stage (3 mm–25 days), there is also a very rapid lengthening of the ileum/colon, that is, the postarterial limb of the midgut and hindgut. The failure of rostro-caudal growth of the URS would cause an arrest of cloacal septation with the hindgut opening above the level of the Wolffian ducts in males and at a similar "high" level in females. The error in mesodermal differentiation in the adjacent hindgut could lead to its "dysmorphogenesis" before it lengthens, ultimately forming the colonic pouch, and also accounts for the high incidence of abnormalities affecting the postarterial limb of the midgut and hindgut.[1,2,4] A variation in cranial extent of the error in mesodermal differentiation in the URS and adjacent hindgut can account for the varying subtypes (Types I–IV) of CPC. Significantly, Wakhlu *et al*.[14] suggested a similar early error in development of the "splanchnic fold" corresponding to the URS. However, in other cases, CPC may be a part of a more generalized, severe mesodermal defect during early embryogenesis.

In girls, the early arrest of cloacal septation with the "high" communication between the colonic pouch and the urinary tract or vestibule, can keep the Müllerian ducts apart during their descent, explaining the double uterus and vagina and stretching out the trigonal area, bladder neck, and urethra.[27,44] Significantly, in males with CPC, where descent of the Müllerian ducts plays no role in the development of the genitourinary tract, the bladder neck is normal.

It is likely that in Type IV CPC associated with a "low" or "intermediate" ARM, a similar but "temporally" later event occurs when cloacal septation has progressed caudally to a greater extent and lengthening of the hindgut has already occurred.

Clinical Presentation and Diagnosis

Males

Newborn males present with absence of the anal opening, often with marked abdominal distension, a tense, shiny abdominal wall, and dilated superficial veins.[1,5,6] Fecaluria may be present in nearly 50% of patients,[1,4] and if severe, abdominal distension may not be marked. Abdominal distension may be less apparent in Types III/IV CPC. In newborns, there is a high incidence of perforation of the thin-walled pouch with pneumoperitoneum, peritonitis, and septicemia.[1,5,6]

Females

Most present with gross abdominal distension as in males. However, others may present later with moderate abdominal distension, poorly managed constipation with episodes of enterocolitis, perineal rashes, and often a doughy feeling over the abdomen due to retained feces.[18,27,44] Urinary incontinence may be apparent.[27,44] Abdominal X-rays in older girls may show a large soft-tissue shadow of retained fecal matter. We believe that examination of the external genitalia, in the clinic/EUA is diagnostic.[27,44] In doubtful cases, a dye-study through the vestibular fistula, if present, may be diagnostic.[2,13]

Radiological Examination

Prone cross-table X-ray/plain X-ray abdomen

The typical feature in Type I or II CPC, seen best on anteroposterior X-rays, is an enormous gas shadow or air-fluid level on the left side of the abdomen, occupying more than 50% of the abdominal width and terminating in a high supralevator position [Figure 8].[1,5,6] The small bowel loops are displaced to the right. The sacrum is usually normal.[2,4] A prone cross-table or lateral film is however, better for visualizing gas within the bladder. In the vast majority, diagnosis is possible by abdominal radiographs alone.[16] The typical X-ray appearance may be absent with a very wide CVF or pneumoperitoneum due to pouch perforation.

In most cases of Type III and IV CPC, a similar but smaller air-fluid level is seen on a plain X-ray, and in the "true" Type IV CPC, often with gas in the bladder [Figure 9]. The dilated colonic loop in Type IV CPC may be more variable in position, perhaps because of greater mobility due to the long sigmoid mesocolon.[45] It is sometimes difficult to differentiate X-ray findings from the terminal rectosigmoid dilatation in ARM; although, the air-fluid level in Type IV CPC is usually better defined.

Management

Types I/II congenital pouch colon

The standard management policy at our institution from 2000 to 2007 was to excise the colonic pouch in all subtypes of CPC[4,31-33,46] as, even after tubularization, the colonic tube has poor peristalsis and emptying along with the risk of significant redilatation.[6,11,46] However, the extremely poor functional results following this management protocol and significantly better results after tubular colorraphy (TC) and pull-through,[47,48] led us to opt for preserving the colonic pouch wherever possible. This has been the prevalent view throughout at other centers.[5,6,11,14,49]

Single versus staged surgery

A single-stage neonatal excision of the colonic pouch/TC and pull-through for all subtypes of CPC[15,50] avoids the complications of diversion procedures, is cost effective, and may have maximum potential for developing normal defecation reflexes.[50] At 3 years

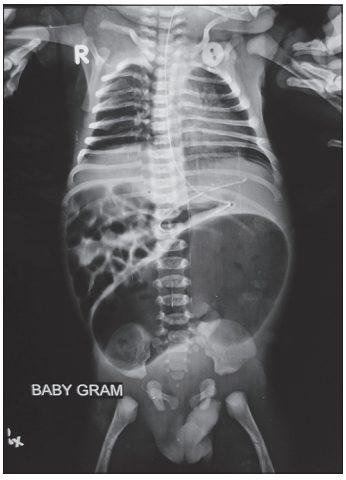

Figure 8: Plain X-ray abdomen in a case of Type I congenital pouch colon showing a massive gas shadow on left side of the abdomen. Note normal sacrum

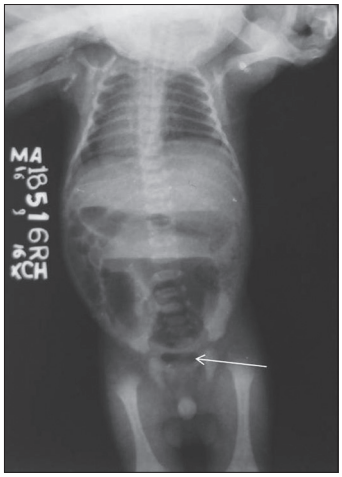

Figure 9: Dilated colonic pouch in "True" Type IV congenital pouch colon. Note gas within the urinary bladder (arrow)

follow-up, Gangopadhyay *et al*.[50] reported good or fair continence in 75% of patients undergoing primary neonatal single-stage procedure as against 48% with staged procedures. However, especially with TC, this is a major surgical undertaking with unacceptably high risks of complications and mortality.[3,11]

Staged repair of Type I/II congenital pouch colon

Primary surgery

Based on recommendations of the IAPS consensus meet,[8] an algorithm for staged management of Types I/II CPC, and some cases of Type III CPC where the normal proximal colon is quite short, is shown in Figure 10. In essence, the colonic pouch should be excised only if it is severely ischemic, gangrenous or has multiple perforations. The diverting options are:

- A loop or divided ileostomy sited sufficiently proximal to the pouch so as not to interfere with either the mobilization or vascularity of the bowel to be pulled-through later.[1,2,4] Complications include major ileostomy losses, failure to thrive, severe peristomal excoriations,[48] and possible metabolic acidosis due to urinary reabsorption through the wide CVF. Use of oral rehydrating solutions and nutritional supplementation are essential[29,48]
- TC and end colostomy[1] wherein the colonic pouch is tailored into a long tube to reduce redundancy, improve peristalsis, and preserve colonic length for its absorptive function. Wakhlu and Wakhlu[49] felt that in newborns, the pouch is thin-walled and the mortality with this procedure is high (30%–50%).[1,5,14] Coloplasty (TC) is easier in a 5–8-month-old baby with a thicker pouch and initially, a window colostomy should be performed.[49] However, in trained hands, the procedure is relatively safe in newborns in good condition and with good pouch anatomy[2-4]

  TC-operative procedure:[49] The approach is by a lower left-quadrant incision, extended downwards and transversely to the right. A similar, smaller incision is adequate for a window-colostomy. The associated fistula is dissected as low as possible and divided between ligatures. While some prefer restricting the reconstructed colonic tube length to 5 cm[50] or 15 cm[11] to reduce complications including colonic redilatation, Wakhlu and Wakhlu[49] advocate making it as long as possible, not >1.5–2 cm diameter, and with an attempt to preserve the tissue of the fistula. This is also our policy
- Window or side colostomy: Although this is a safe, short, easily performed surgery, it is very frequently associated with stomal stenosis,[1,5,25,37] prolapse, often with eversion of the entire colonic wall,[1,2,4,5,17,32,37] and poor functioning and emptying necessitating

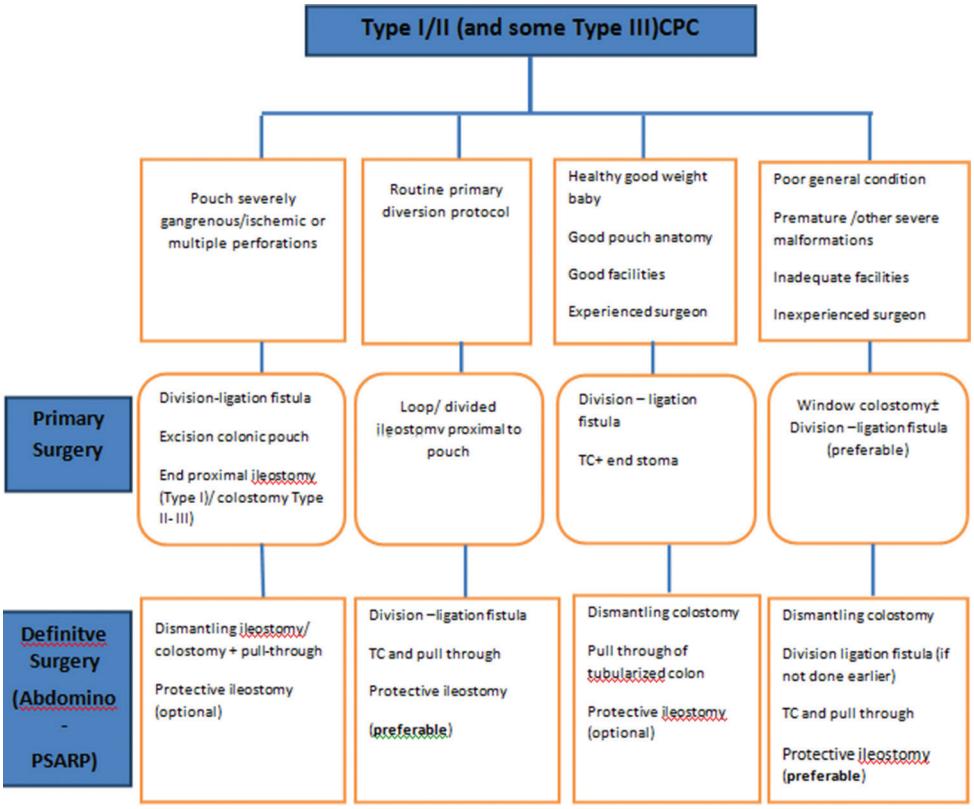

Figure 10: Algorithm for management of Types I–II congenital pouch colon

regular washouts.[15,17,25,37] There is also a failure to thrive and a mortality rate of 15%–20%.[16,49] Urinary reabsorption can result in metabolic acidosis. A preliminary window colostomy still has a role in certain situations [Figure 10]. If feasible, division-ligation of the CVF avoids persistent fecal contamination of the urinary tract. It should also be sited [Figure 2] away from the segment of the pouch that will be tubularized later.

Definitive surgery

This is usually performed at the age of 4–18 months. If a window colostomy/an ileostomy had been performed initially, early surgery is necessary to minimize stomal complications as well as those due to the wide CVF. Mandatory preoperative investigations include an abdominal ultrasound and considering the high incidence of VUR, a voiding cystourethrogram (VCUG). A spiral computed tomography scan with three-dimensional reconstruction of pelvic musculature[34] or a pelvic magnetic resonance imaging scan is optional.[3]

The posterior sagittal approach along with laparotomy (Abdomino- PSARP) offers excellent exposure for accurate placement of the bowel.[1,2] Recently, laparoscopic- assisted anorectoplasty (LAARP) for Type III CPC has been described,[19,51] the entire procedure being carried out with the patient in the supine-lithotomy position. A similar procedure can be performed with laparotomy, avoiding changes in the patient's position. Occasionally, a previously tapered pouch may have redilated,[1,2] necessitating its retubularization. A protective ileostomy is preferable in cases where TC is performed during the definitive procedure and can be closed 3–6 months later. A regime of daily anal dilatations is begun 3 weeks after pull-through.

Staged repair of Type iii and iv congenital pouch colon

Primary surgery

For Types III/IV CPC, the best primary option is a diverting colostomy quite proximal to the colonic pouch.

Definitive surgery

Preoperatively, after confirmation by a distal colostogram, "True" Type IV CPC with a CVF mandates an abdomino-PSARP for ligation of the CVF. We believe, as do others,[49] that the terminal bowel (recto-sigmoid) should be preserved, usually with tubularization of the dilated segment. This is brought down to the perineum in the 2nd stage and the protective colostomy closed later. In recent years (2007–2015), we have performed this procedure successfully in 6 boys with "True" Type IV CPC.

In Type III/IV CPC with a relatively low termination, in the first stage (PSARP), the distal colonic pouch is mobilized, tapered and brought to the perineum. Three to 6 months later, trans-abdominal tubularization of the dilated colon is performed with closure of the colostomy preferably at the same stage, or subsequently. A similar procedure for Type IV CPC with a low ARM was described by Atabek *et al*.[20] An alternative in the second stage of definitive surgery is the procedure employed for the management of megasigmoid associated with repaired ARMs[21,22] in which subtotal resection of the colonic pouch is performed with anastomosis of normal proximal colon to the distal remnant of the colonic pouch.[4]

Many,[3,11,12,23,50] however, believe that as the length of normal proximal colon is sufficient for absorptive function, the best primary surgical option is excision of the colonic pouch with an end-colostomy followed later by pull through of the stoma site.

Management of Congenital Pouch Colon in Girls

The principles of management are similar to those in males. A VCUG and preoperative cystourethro-genitoscopy are essential.[44] The management of urinary incontinence may need bladder neck repair (BNR).[27,44] We performed, Young-Dees BNR in 6 girls with urinary incontinence, with moderate improvement in 2 (dry interval up to 3 h) and little or no improvement in 4 girls. A part of the colonic pouch could be preserved for possible use for bladder augmentation.[44] Monitoring for the obstetric implications of the associated genital anomalies is a long-term requirement.[27,44]

Girls with CPC do not have a "persistent cloaca" in the accepted sense.[27,44] Extensive surgeries for the vaginal component of the abnormality such as total urogenital mobilization are not required as there is a wide vestibule and introitus.[27,44] Division of the often thick intervaginal septum and vagino-pouch remnant fistula on either side (if VF present),[28,44] can produce a single capacious cavity with a relatively normal/hypospadic urethral opening.[44]

Complications after definitive surgery

Suture-line leak after TC with its consequences and wound infection and/or dehiscence, although uncommon, are the major short-term complications[16] and can be minimized by a protective loop ileostomy.[3,11,16] Mucosal prolapse, anal stenosis, and skin excoriations are other frequent complications.[11,16]

Massive redilatation of the tubularized colon after TC and pull-through in Types I/II CPC is

troublesome.[6,11,47] Its incidence can be minimized by constructing a narrow (1.5–2 cm diameter) colonic tube initially.[49] Pharmacological manipulation and aggressive bowel management with washouts can also minimize this risk. In some cases, sub-total excision of the redilated pouch may be necessary.[46]

Results of Treatment and Long-term Follow-up

The mortality rate in CPC has reduced from around 40%[16] to 15% with increased awareness of the condition and improvement in care and management.[3,11]

Most long-term follow-up studies report fairly good results regarding both fecal continence and growth and development.[5,6,15,49,50] Our results are not so encouraging. Puri *et al*.[47] found that all 13 patients with an ileal pull-through had height and weight less than the 50th percentile of the expected value for age. Nine patients with Types III/IV CPC or Type I/II CPC with pull-through after TC had near-normal growth patterns or parameters between the 50th and 80th percentile.[47] A more recent study from our center of 31 patients, at least 3 years old, showed severe fecal incontinence with continence score[52] of 5 or less in 20 patients (64.52%).[48] The remaining 11 patients (25.81%), including the 10 patients with Types III/IV CPC, had "poor" fecal continence.[52] There was a high incidence of malnutrition, especially "stunting" (low height-for-age) which was the most common in the 0–5 years age-group.[48] Patients with Types I/II CPC had a higher incidence of anemia and severe fecal incontinence than those with Types III/IV CPC. Patients with Types I/II CPC, managed by excision of the colonic pouch had a higher incidence of severe fecal incontinence and malnutrition than those with pull-through after TC, suggesting that the latter procedure has long-term benefit.[48] However, TC and pull-through is associated with recurrent abdominal distension and frequent attacks of enterocolitis, necessitating frequent need for antibiotics and colonic washouts.[47] Patients with CPC, especially its more severe forms, also need monitoring of the nutritional status.[48] Dietary supplementation of micronutrients, and early institution of high energy, good-quality protein intake are necessary.[48]

Managing fecal incontinence in Types I/II CPC patients is very challenging. In the immediate postoperative period, children usually pass liquid stools very frequently, often more than 20 times per day. Perineal excoriations with secondary bacterial and fungal infection and formation of granulomas are very common.[4] Oral loperamide/diphenoxylate,[48] and twice-daily colonic washouts (12.5 ml/kg body weight per dose) may improve the continence status, although the dry interval is not sufficient. Although stool frequency and consistency usually improves over several months, the quality of life of a patient of Type I/II CPC with severe fecal incontinence and perineal excoriations is very poor and a permanent abdominal end-colostomy following TC, with an abdominal stoma bag, may well be a better option.[4]

Future directions

In the recent past, several authors[3,30-32] have detailed the abnormalities found on histological examination of the colonic pouch. However, there is still a need to establish a histopathological correlation with the similarity between all subtypes of CPC as well as with megarectum-megasigmoid associated with ARM. The specific etiopathogenetic factor or teratogen responsible for the occurrence of CPC is obviously difficult to identify. Recently, Maudar *et al*.[53] reported an immunohistochemical study of colonic pouch tissue that revealed an enhanced expression of beta-catenin, Ihh, Notch1, HMGA1 and commented on the expression of molecules from all three embryonic signaling pathways. Future studies may help elucidate the exact signaling pathways and gene expressions that are altered in CPC as compared to those in the usual ARM.

Detailed and comprehensive follow-up studies carried on till adulthood are also necessary to properly evaluate the results of various management protocols. The anomalous anatomy of the genitourinary tract in girls, as described here and earlier,[27,44] needs wider acceptance. Managing urinary incontinence in girls is a difficult task and more centers need to report the procedures used and results achieved in managing this problem. As of now, the lot of the girl with CPC is particularly unfortunate, having to manage both urinary incontinence and the very frequent severe fecal incontinence.

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Conflicts of interest

There are no conflicts of interest.

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