Congenital Pouch Colon Revisited

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Background/Purpose: The authors' recent experience with the study of the presentation, the pathological anatomy, and results of management of congenital pouch colon (CPC) malformations is presented. The possible embryogenesis of this condition is discussed.

Methods: Between January 1991 and June 1997, CPC with anorectal agenesis was diagnosed in 39 patients, 27 boys and 12 girls, who were classified in four groups, based on the length of the normal colon proximal to the distended segment. In 31 patients with little or no normal colon, the choice of primary procedure was based on the condition of the infant and the viability of the colonic pouch. Definitive surgery has been performed in 15 patients from this group. The eight patients with a suitable length of normal colon had a lower level of termination of the colonic pouch and a lower fistula. In these, a colostomy was constructed just proximal to the pouch, with later definitive surgery in four patients consisting of excision of the colonic pouch and pull-through of the proximal colon.

Results: Mortality after primary surgery was 13%. Definitive surgery was well tolerated in all 19 patients.

Conclusion: In patients in whom a tubularized segment of the colonic pouch was used, continence was only fair to poor a year later.

INDEX WORDS: Pouch colon, congenital; short colon, congenital; anorectal malformations.

CONGENITAL POUCH COLON (CPC), also referred to as congenital short colon[1-4] or "pouch colon syndrome"[5] is an unusual malformation in which a globular dilatation of a shortened colon is associated with an anorectal malformation (ARM), usually of the high variety. Most large series describing this condition have been reported from Northern India,[5-8] and several recent studies[5,7,8] have elaborated on the presentation, the pathological anatomy, and management of CPC. The present report outlines our recent experience with the management and follow-up of patients with a pouch colon malformation. Recent observations that patients with a reasonable length of normal colon appear to have a lower level of termination of the colonic pouch and a lower fistula have led us to renewed speculation on the embryology of this condition and its associated malformations.

MATERIALS AND METHODS

From January 1991 to June 1997, 598 patients with anorectal malformations (ARMs) were treated in the Department of Pediatric Surgery, Kalawati Saran Children's Hospital, New Delhi, India. Thirty-nine of these patients (6.5%) were found to have an associated CPC malformation.

Twenty-seven of the patients were boys and 12 were girls (male to female ratio, 2.25:1). All the boys and seven of the girls presented in the neonatal period; the other five girls with a nearly normal length of colon and a vestibular fistula received diagnosis between the ages of 3 and 8 months.

In this study, a modification of the classification described by Narasimharao et al[5] is used; the patients categorized into four types (Fig 1) as follows: type I, normal colon is absent and the ileum opens directly into the colonic pouch; type II, the cecum along with a segment of normal colon (up to around 8 cm in the newborn) is present proximal to the colonic pouch; type III, the normal colon proximal to the colonic pouch extends at least to the level of the hepatic flexure but does not extend beyond the descending colon; type IV, the colon is of nearly normal length, only the terminal portion (rectum and a varying length of the sigmoid) is converted into a pouch.

The incidence of the various types of CPC is shown in Table 1. In the four girls with type I or II CPC and persistent cloaca, there was a urogenital sinus malformation, and the colonic pouch terminated in a fistula to the posterior wall of the bladder. In addition, a septate vagina was present in one of these patients. A completely bifid uterus and cervix along with a septate vagina was present in another girl with persistent cloaca as well as in two girls with a type I or II CPC and colovesical fistula and the patient with a type II CPC and vestibular fistula.

In both girls with a type III CPC and vestibular fistula, the normal proximal colon extended to the splenic flexure.

The associated malformations are shown in Table 2. Two boys with a type II CPC had duplication of the appendix as well as a Meckel's diverticulum 3 to 5 cm proximal to the ileocecal junction, indicating shortening of the terminal (or distal) ileum.

On the basis of the findings of the invertogram, the three boys with type IV CPC were thought to have a supralevator anorectal anomaly, with only one having the large air-fluid level suggestive of CPC. In the five girls with a vestibular fistula, the type III or IV CPC was detected only during preliminary surgery; all of these patients had presented with poorly managed constipation, and two girls also had recurrent abdominal distension with overflow incontinence.

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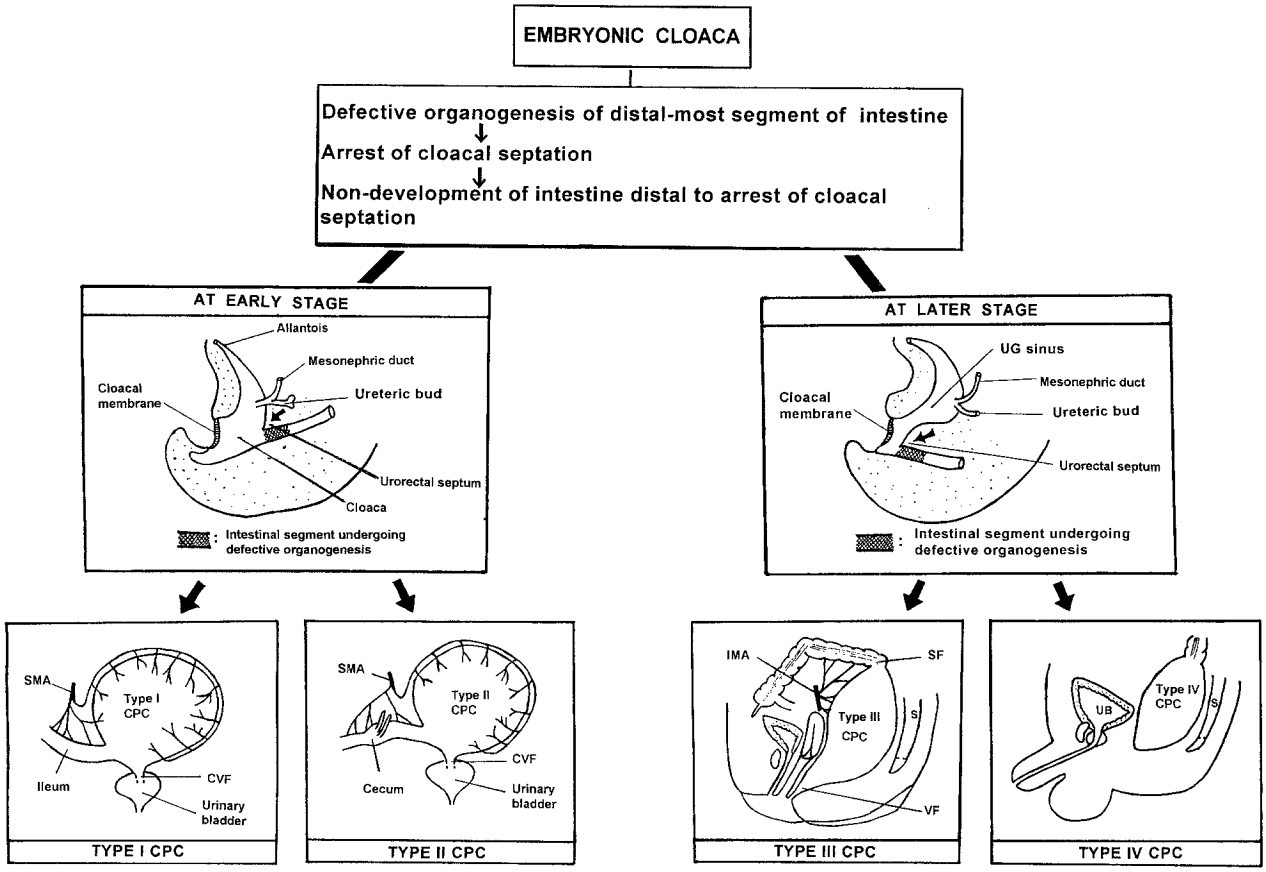

Fig 1. Diagram shows the likely embryogenesis of the four types of congenital pouch colon. CPC, congenital pouch colon; SMA, superior mesenteric artery; IMA, inferior mesenteric artery; CVF, colovesical fistula; SF, splenic flexure; VF, vestibular fistula; UB, urinary bladder; S, Sacrum.

Primary Operation

In patients with type I or II CPC, the choice of primary procedure was based on the protocol previously described.[7] In one newborn girl with a type II CPC and vestibular fistula, during primary surgery, the colonic pouch was mobilized as far down as possible, divided at this point, and subtotal excision of the pouch with tubular colorrhaphy performed. We plan to excise the distal remnant of the colonic pouch and vestibular fistula at the time of pull-through of the tubularized colonic segment during definitive surgery.

Table 1. Incidence of Subtypes of Pouch Colon Malformation

Type	No. of Boys	No. of Girls
I	10 (all with colo-vesical fistula)	3 (2 persistent cloaca) (1 colovesical fistula)
II	14 (all with colo-vesical fistula)	4 (2 persistent cloaca) (1 colovesical fistula) (1 colovestibular fistula)
III	—	2 (all with coloves-tibular fistula)
IV	3 (2 blind-ending pouch at level of prostate). (1 colobulbar-urethral fistula)	3 (all with coloves-tibular fistula)

In the eight patients with a type III or IV CPC, a divided transverse or descending colostomy was performed just proximal to the colonic pouch. An attempt to excise the colonic pouch by the abdominal route was unsuccessful in one patient (a boy with a type IV CPC);

Table 2. Associated Malformations

Type	Malformation	No. of Patients
I	Gastrointestinal tract	
	Double appendix	6
	Tiny appendix	4
	Absent appendix	6
	Meckel's diverticulum	2
	High jejunal atresia	1
II	Genitourinary tract	
	Vesicoureteric reflux	15
	Unilateral renal agenesis	4
	Hydronephrosis	4
	Cloacal Anomaly	4
	Septate vagina	1
	Completely bifid uterus and cervix with septate vagina	4
III	Cardiovascular system	
	Cyanotic congenital heart disease	1
IV	Skeletal system	
	Partial sacral agenesis	2

subtotal excision of the pouch was performed, the distal remnant closed, and a proximal descending end-colostomy constructed. Subsequent retrograde urethrogram (RGU) showed a fistula from the terminal portion of the pouch remnant to the bulbar urethra.

Definitive Operation

The definitive operation is performed when the child is thriving, generally at 4 to 18 months of age. Nineteen of the 39 patients have undergone reconstruction (including two girls in whom one-stage reconstruction of a persistent cloaca was possible). In all cases with type I or II anomalies, the posterior sagittal approach[9] was used, the additional abdominal component being mandatory with this anomaly. The details of the surgical procedure were described in our previous report.[7]

In patients with type III or IV CPC, the posterior sagittal approach was used to mobilize the distal colonic pouch from its vestibular opening in girls and its lower limit in boys, carrying the dissection as high as technically possible. A wide-bore rubber tube was placed through the pelvis after completion of this dissection. During the abdominal phase of the surgery, the colon between the colostomy stoma and the previously mobilized lower end of the colonic pouch was excised. The proximal colon was mobilized, tapered if necessary, sutured to the rubber tube, and brought down to the site of the neoanus. A protective ileostomy (type III) or transverse colostomy (type IV) was performed.

From January 1986 to June 1997, 80 patients with CPC have been treated at our institution. This includes 41 patients described in our previous report[7] and the 39 patients in the present series. Of these 80 patients, 25 (all with type I or II CPC) have undergone definitive surgery, which included the pull-through of a tubularized segment of the colonic pouch. These include 13 patients from our previous report[7] and 12 patients from the present series. Of 18 patients in whom all surgeries, including closure of the protective loop ileostomy, had been completed a year or more before the study, only seven returned for physical examination, barium enema, and assessment of fecal continence and bowel function as described by Kiesewetter and Chang.[10] None had sacral abnormalities or evidence of neuropathic bladder dysfunction.

RESULTS

The primary operative procedures performed on the 39 patients are shown in Table 3. Five postoperative deaths (13% mortality rate) resulted from (1) bronchopneumonia and septicemia in a child in whom a thin-walled gangrenous type I colonic pouch was excised and an end-ileostomy performed; (2) an anastomotic leak in two patients with peritonitis after tubular colorrhaphy; (3) prematurity (28 weeks) with a high jejunal atresia and cyanotic congenital heart disease in a boy with type IV CPC; and (4) poor general condition, dehydration, and septicemia after window colostomy in a child with a type II CPC.

Histological examination of the tissue resected from the colonic pouch in 25 patients showed normal colonic mucosa and ganglion cells. Thinning of the muscle layers was noted in 12 specimens, all from infants who underwent surgery in the neonatal period, whereas hypertrophy of the muscle layers was present in the excised colonic pouch obtained from two girls with a type III and IV CPC and vestibular fistula.

The definitive operative procedures performed in 19 patients are shown in Table 4. Postoperative complications include one death related to blood transfusion, one wound infection with retraction of the pulled-through colon, and two minor wound infections.

Table 3. Primary Operative Procedure

Type of Malformation	Procedure	No. of Patients	Postoperative Mortality Rate (No.)
Type I & II (n = 31)	Window colostomy alone	2	1
	Excision of gangrenous thin-walled pouch with end ileostomy	4	1
	Excision of pouch with end colostomy	3	—
	Disconnection of fistula with end colostomy	3	—
	Fistula disconnection, subtotal excision of pouch and tubular colorrhaphy	19	2
Type III (n = 2 girls with vestibular fistula)	Divided transverse colostomy proximal to pouch	2	—
Type IV (n = 6)	Divided descending colostomy proximal to pouch	5	1
	Subtotal excision of pouch, descending colostomy proximal to pouch	1	—

Seven patients who had undergone pull-through after tubular colorrhaphy were assessed for continence by the Kiesewetter and Chang method.[10] The result was only "fair" in two patients and "poor" in five others who had frequent fecal soiling day and night. They all had semisolid stools, four with frequent episodes of diarrhea. Barium enema results showed the caliber of the colonic tube to be within normal limits in five patients and moderately dilated in two children with poor continence (Fig 2). Twice-daily tap water enemas (12.5 mL/kg body weight per dose) improved the continence status in three patients with poor fecal continence, but the dry interval after administration of the enema was only 4 to 8 hours.

DISCUSSION

A high terminal fistula with the genitourinary tract previously has been regarded as a characteristic feature of all four types of CPC associated with anorectal agenesis.[5,7,11] However, in one report, 10 of the 108 patients with CPC also had a colourethral or colovestibular fistula, but the actual length of normal proximal colon was not noted.[8] A recent study[12] as well as another report (Krishna A, personal communication, May 1997) also indicate that type III or IV CPC is associated with a lower level of termination of the colonic pouch.

Table 4. Definitive Operative Procedures (19 Patients)

Status at Time of Definitive Operation	Definitive Operation (Combined Posterior Sagittal and Abdominal Approach)
Tubular colorrhaphy and end colostomy (n = 13) (Type I/II anomaly)	Disconnection of end colostomy; pull-through of previously tapered colon with protective ileostomy (n = 6) Retubularization to tube of 2.5 cm diameter; pull-through of retubularized colon with protective ileostomy (n = 4) Excision of redilated colonic pouch; pull-through of proximal colon after tapering; protective ileostomy (n = 2) Excision of small redilated pouch; pull-through of proximal ileum (n = 1)
Window colostomy without disconnection of fistula (n = 1) (Type II anomaly)	Disconnection of fistula, subtotal excision of pouch with tubular colorrhaphy; pull-through of tubularized colon with protective ileostomy
Fistula disconnection with end colostomy (n = 1) (Type II anomaly)	Subtotal excision of pouch with tubular colorrhaphy; pull-through of tubularized colon with protective ileostomy
Divided colostomy proximal to pouch (n = 2 girls with Type III anomaly and vestibular fistula; 2 Type IV anomaly)	Excision of pouch from distal colostomy stoma to lower limit of pouch; pull-through of proximal colon with protective ileostomy (2, Type III) or transverse colostomy (2, Type IV)

Patients with type III or IV CPC constitute only a small percentage of the cases reported in large series,[5-8] and it is possible that in these patients, the anatomy of the colon and the colonic pouch was not studied very well. Although the differentiation between types II and III CPC may on occasion be subjective, the classification presented here appears to provide a clear distinction, the pouch ending with a high fistula in types I and II and a low fistula in types III and IV CPC. There may be occasional exceptions; the patient reported by El Shafie[13] had a type II CPC with a perineal fistula, whereas one of our patients had a type II CPC and vestibular fistula.

The following discussion on the probable embryopathogenesis of CPC is based on a study of the large number of patients described in several series, including our own reports.[1,5-8,11]

Certain features characteristic of type I or II CPC are also seen in cloacal exstrophy, and this has been noted previously.[3,14] In both these conditions, there appears to be an arrest at an early stage in partitioning of the cloaca by the urorectal septum.[1,11,15-17] With CPC, it is likely that the primary abnormality is "defective organogenesis" or "primary dysplasia" of the distal-most segment of intestine close to the descending urorectal fold (Fig 1). This defective segment could anchor the entire cloaca ventrally and prevent further descent of the urorectal septum (Spencer R, personal communication, June 1997). During later development, the abnormal segment would dilate to form the colonic pouch with its unique anatomic and physiological characteristics.[7] These characteristics are very similar to those of the dilated colonic segment in patients with segmental dilatation of the colon (SDC)[18-20] and significantly, "impairment of intestinal organogenesis"[21] and "primary dysplasia"[22] have been suggested as the etiological factors responsible for segmental dilatation of the intestine.

In both cloacal exstrophy and type I or II CPC, the early arrest of cloacal septation results in imperforate anus, appendiceal abnormalities,[1,5-8,17] and appears to interfere with the longitudinal growth of the hindgut.[1,14] The distal colon and rectum are absent[1,5-8,17] and the inferior mesenteric artery is usually missing.[1,5,7,11,17] As shown by the frequent additional presence of a Meckel's diverticulum in type I or II CPC[3,7] as well as in cloacal exstrophy (Spencer R, personal communication, June 1997), often with shortening of the ileum distal to the diverticulum, this shortening of the intestine seems to involve most of the bowel caudal to the omphalomesenteric duct.

With type I or II CPC, the early arrest of cloacal

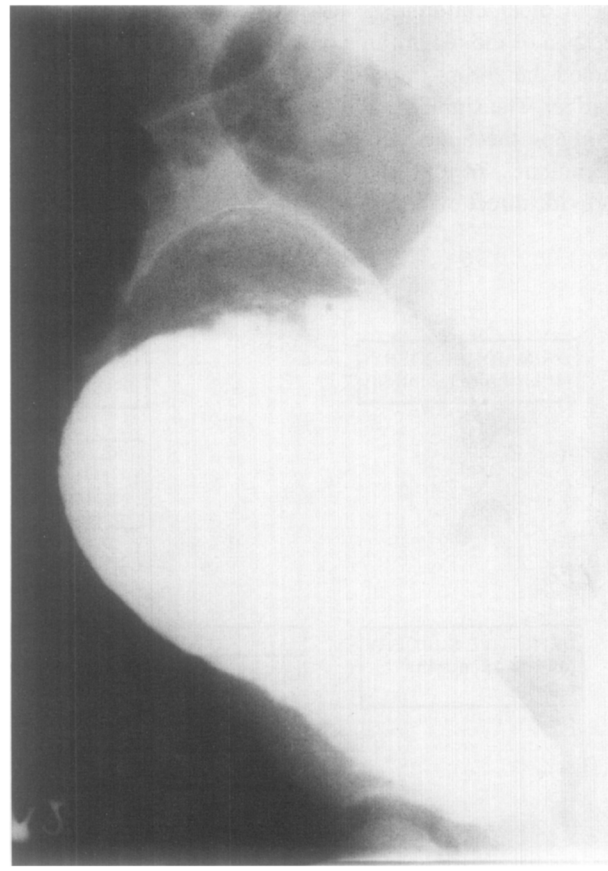

Fig 2. Barium enema findings in a child with moderately redilated colonic tube after pull-through after tubular colorrhaphy.

septation can account for the high and often wide fistula with the genitourinary tract.[1,7,11] Relative delay in the event of "defective organogenesis" and the consequent arrest of cloacal septation would result in type III or IV CPC with a corresponding longer length of normal proximal colon and a lower fistula (Fig 1).

The classical theories of cloacal partitioning[23,24] have been challenged by Van der Putte[25] and Kluth et al.[26] However, these newer theories[25,26] cannot explain some of the major findings seen in cloacal exstrophy and type I or II CPC. If a long segment of distal midgut and the hindgut is derived from differentiation of the cloaca distal to the allantois, partitioning of the cloaca by the urorectal fold should be "real" rather than just "apparent" as suggested by Kluth et al.[26]

Significantly, CPC is infrequently associated with major malformations in other organ systems.[1,7] Despite the high nature of the ARM associated with type I or II CPC, major sacral abnormalities are rare,[8,11] and the voluntary musculature of the pelvis and perineum is usually well developed.

It is evident that type IV CPC as described by us is very similar to the terminal rectal or rectosigmoid dilatation reported in children with repaired ARMs, usually of the low variety.[27-30] Although most of these patients have been older children,[27-30] Cloutier et al[28] reported focal ectasia of the rectum in a newborn with a low ARM and noted the abrupt transition to proximal colon of normal caliber. The similarity with SDC also been has noted.[28,30] Perhaps the application of sophisticated histochemical techniques in the study of resected specimens may provide direct evidence of a related origin and a common abnormality in all four types of CPC, megarectum-megasigmoid as well as SDC.

In a previous report,[7] we detailed a protocol for the management of the various types of CPC. However, a recent report[31] has confirmed our observation that the colonic pouch is inherently abnormal and, even after tubular colorrhaphy, may resume its characteristic dilatation, although variable in degree, over a period of time. In 4 of the 13 patients (30%) in the present series in whom tubular colorrhaphy had been performed during primary surgery, redilatation of the colonic pouch necessitated retubularization during definitive surgery. However, we believe that in type I or II CPC, wherever possible, tubular colorrhaphy should still be performed to preserve the absorptive function of a relatively long segment of colon, but in patients with type III or IV anomalies, the length of normal colon proximal to the pouch is sufficient for this function. The protocol we have followed in managing type III or IV CPC is shown as Plan A in Fig 3.

Bowel function and continence have not been evaluated yet in our patients with type III or IV anomalies in whom the colonic pouch was excized. As stated earlier, type III and especially type IV CPC is very similar to the megarectum-megasigmoid reported in some patients with ARMs.[27-30] It is possible that even in type III or IV CPC, internal sphincter tissue and sensory and proprioreceptors may be present in the terminal bowel. The importance of preserving the distal rectum for its reservoir function also has been stressed.[30] A modification of the surgical procedure successfully employed for the management of megasigmoid associated with ARMs[29,30] may be an appropriate alternative method for treating

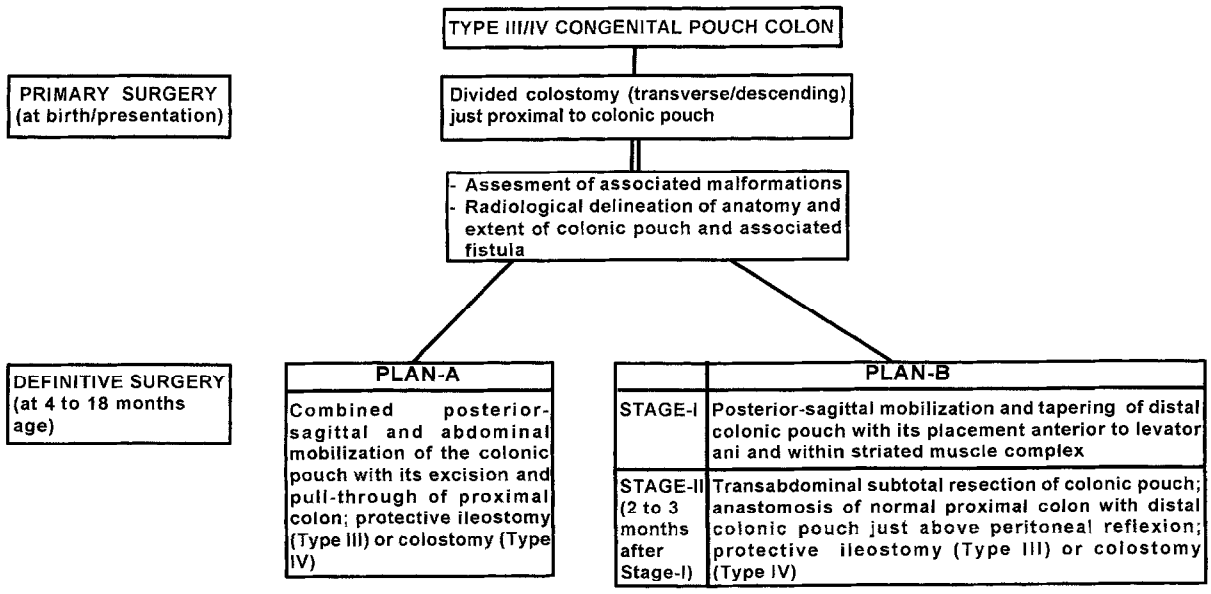

Fig 3. Suggested management options (PLAN A and B) for type III or IV CPC.

these patients (Plan B in Fig 3). Currently, we plan to use this protocol (Plan B) for managing type III or IV CPC so that later we can compare the results achieved with these two contrasting modes of management.

Assessment of bowel function and continence in patients with type I or II CPC who have undergone pull-through after tubular colorrhaphy has been possible only in seven patients, all under the age of 5 years, and the results are not very encouraging. In contrast, a similar study reported socially acceptable fecal continence in patients followed up for up to 8 years after pull-through after tubular colorrhaphy.[31] It is possible, therefore, that continence levels in these patients may improve with time. However, complete absence of normal distal bowel, shortening of the overall length of the intestine, as well as absence of the ileocecal valve (with type I CPC) suggests that irrespective of the type of surgical procedure performed, overall fecal continence levels in these patients may not be very good.

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