original article

Congenital pouch colon syndrome: A report of 17 cases

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BACKGROUND: Congenital pouch syndrome (CPC) is a rare condition seen in association with anorectal malformation that occurs almost exclusively in northern India. We reviewed cases seen in our institution to study aspects of clinical presentation, diagnosis, embryogenesis and management and raise awareness of this relatively infrequent entity.

PATIENTS AND METHODS: From March 2002 to September 2004, 17 neonates/infants (11 males and 6 females) treated for CPC associated with anorectal malformations included 13 with type IV and 4 with type I CPC. Diagnosis was made by a single large air-fluid level on the infantogram occupying more than 50% of the entire abdominal dimension.

RESULTS: In all patients, the pouch had fistulous communication with the genitourinary system, and there were other associated anomalies as well. Of 13 patients with pouch colon type IV, 11 neonates underwent laparotomy, ligation of the fistula, excision of the colonic pouch and end colostomy as a stage 1 procedure. Subsequently, these patients underwent definitive surgery, i.e. abdominoperineal posterior sagittal anorectoplasty (AP-PSARP), with or without covering colostomy. Two of 4 patients with type I CPC underwent laparotomy, ligation of the fistula and colorrhaphy as a first-stage operation before AP-PSARP. In our series, 4 patients were diagnosed intraoperatively and were treated in accordance with their operative findings. Post-operatively, there were no major complications except wound infection in some patients. There was one death that was not related to surgery.

CONCLUSION: There are variants of the anomaly, but the possibility of CPC needs to be kept in mind as a possible association with anorectal malformations.

Congenital pouch colon (CPC), also known as congenital short colon,[1,2,3,4] or "pouch colon syndrome" is a rare condition that occurs in association with anorectal malformations. In this condition, the colon is either partially or completely replaced by a pouch-like dilatation communicating with the urogenital tract by means of a fistula (Figure 1). This anomaly is exclusively seen in Northern India, hence all reported series are from this part of world,[4,5,6,7] with only a few cases reported from elsewhere.[9,10,18] It has been classified into four different types depending on the extent of colonic involvement.[6] Frequent associated malformations include the genitourinary and gastrointestinal systems and vertebral anomalies. Management of CPC varies from simple excision of the pouch to colorrhaphy before definitive abdominoperineal pull-through.

PATIENTS AND METHODS

From March 2002 to September 2004,[17] neonates with CPC associated with anorectal malformation were treated in the Department of Pediatric Surgery, Sher-i-Kashmir Institute of Medical Sciences, Soura, Srinagar Kashmir, India. The anatomical classification described by Narasimharao et al[6] was used to categorize these patients as type I (normal colon absent, ileum opens directly into colonic pouch), type II (cecum along with a short segment of colon present proximal to colon pouch), type III (normal colon proximal to pouch extending at least to the level of hepatic flexure, but not beyond descending colon), and type IV (colon of nearly normal length, only rectum and varying length of sigmoid colon replaced by the pouch [Figure 2]).

In our series, we found only two types with type IV CPC in 13 patients and type I in the remaining 4 neonates. There were 11 males and 6 females. These patients presented in the first 72 hours of life with anorectal malformations. Eleven patients of type IV CPC and 2 patients of type I were diagnosed at the time of admission by a single air fluid level seen both on infantogram and invertogram, occupying more than 50% of entire abdominal cavity (Figure 3). These 11 patients with type IV CPC underwent ligation of the fistulous com-

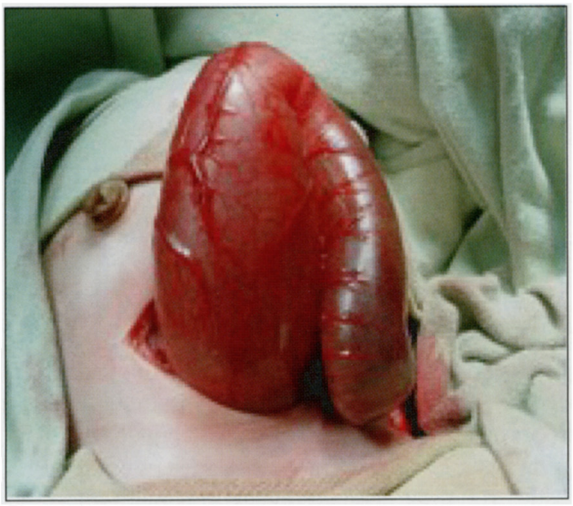

Figure 1. Partial colonic dilatation (type IV)

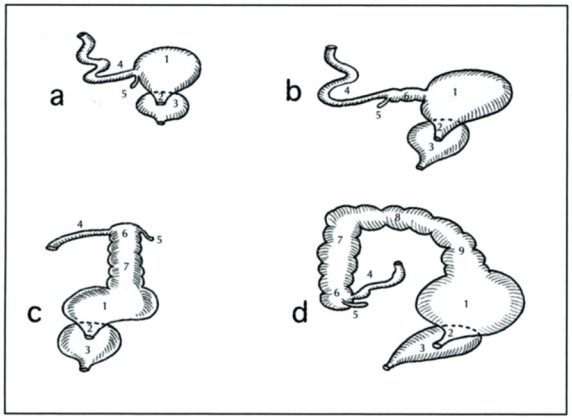

Figure 2. Types of congenital pouch colon. (a) Type I, (b) Type II (c) Type III, (d) Type IV. (1) Colonic pouch, (2) fistula, (3) bladder/vaginal, (4) ileum, (5) appendix, (6) cecum, (7) ascending colon, (8) transverse colom (9) descending colon.

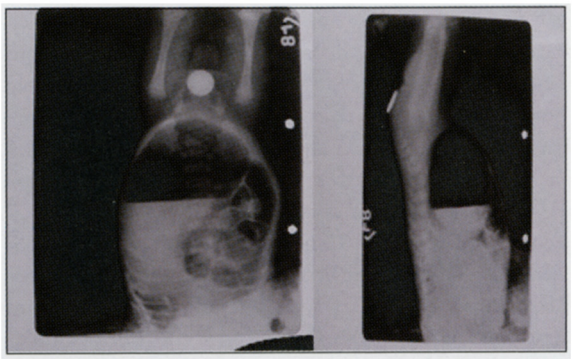

Figure 3. Anterioposterior and lateral view inventogram showing distended colonic pouch occupying more than 50% of the width of the abdomen.

munication with the genitourinary system with primary excision of the pouch and the end colostomy as a stage 1 operation. The definitive, abdominoperineal posterior sagittal anorectoplasty (AP-PSARP) was done at an average age of 9 months to 1 year, with a covering colostomy in 5 patients that was closed 3 to 6 months later. The twelfth patient with type IV CPC underwent a window colostomy that retracted in the immediate post-operative period. This patient was discovered to have a pouch on emergency laparotomy; the pouch was excised after ligating its fistulous communication with the bladder; the AP-PSARP was performed in the same sitting, with a covering colostomy. The one remaining patient with type IV CPC was diagnosed only at the time of definitive surgery; in this patient, a colostomy was made in the sigmoid colon without recognition of the pouch at the time of presentation. At that time, the colostomy was dismantled as the colonic length between the colostomy and the pouch was too short for pull-through; the pouch was excised after fistulous ligation and AP-PSARP was performed in the same sitting.

In 2 of 4 patients with type I CPC, the management included division of the fistula, colorrhaphy and colostomy as a stage 1 operation, AP-PSARP as stage 2 and colostomy closure as a stage 3 operation. The third case, which was operated at another hospital, had bilateral flanks stomae. Laparotomy revealed a type I pouch that required colorrhaphy before definitive surgery. The remaining child with type I CPC had a very small pouch colon with an ill-defined and precarious blood supply.

RESULTS

In all of our patients, the CPC was associated with a variety of anorectal malformations and presented with a blind perineum. All except two patients had severe abdominal distension; these two patients had a large fistulous communication with the bladder causing meconuria. The primary procedure was laparotomy followed by excision of the pouch and end-colostomy in 11 patients with type IV CPC and colorrhaphy and diversion in 2 patients of type I CPC. One patient with type IV who was missed at initial presentation had undergone window colostomy that retracted in the immediate post-operative period. Although he was taken for definitive surgery, the infant died after a month because of septicemia that was not related to a surgical cause. The vascular supply of the pouch was abnormal, with absence of an inferior mesenteric artery and a terminal branch of the superior mesenteric artery supplying the pouch. In one patient, the pouch was supplied by a branch artery in the small bowel mesentery and when mobilized the pouch became gangrenous and required

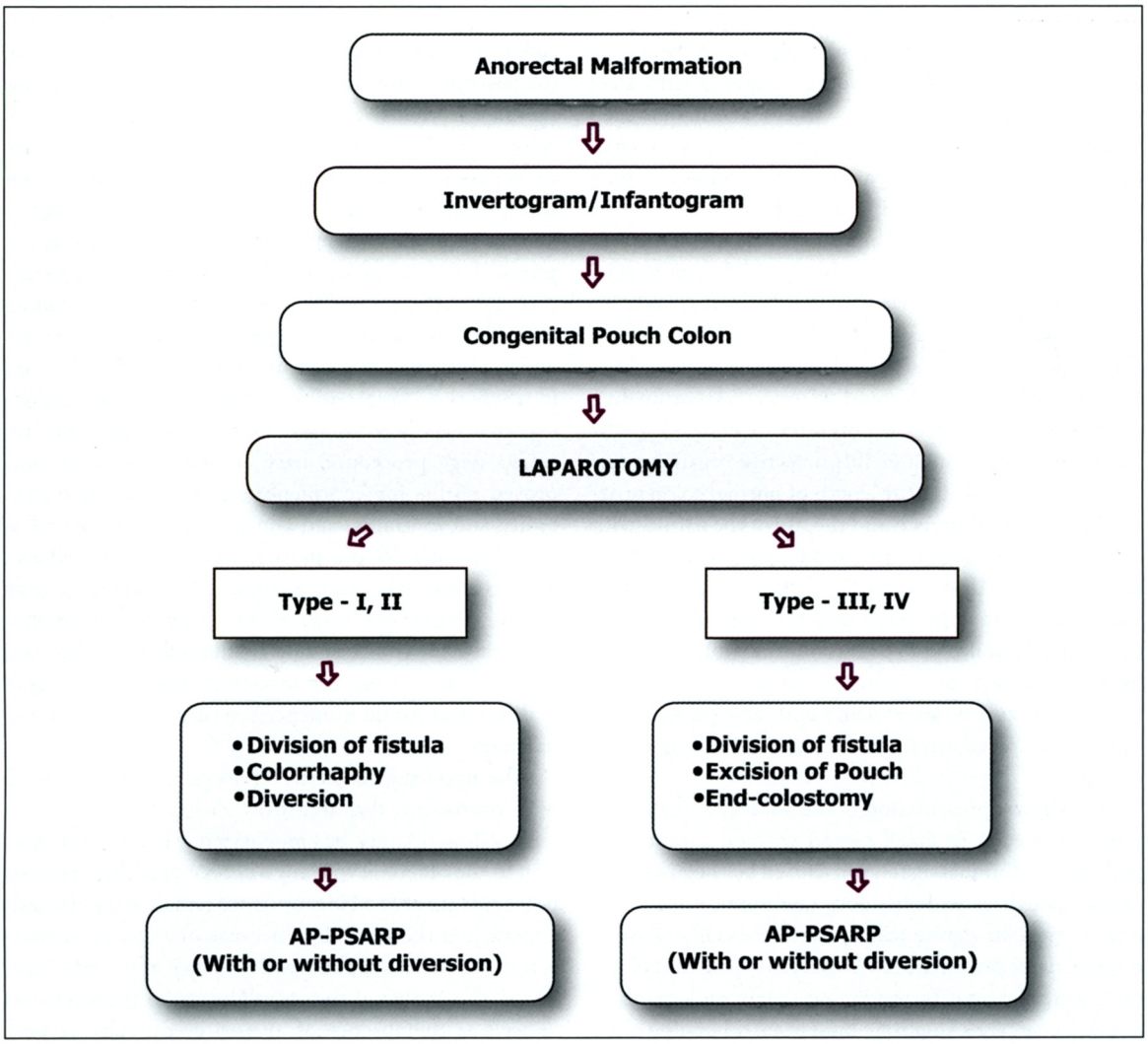

Figure 4. Algorithm for the management of congenital pouch syndrome

excision and end ileostomy. Histological examination of the resected tissue from the colonic pouch showed normal colonic mucosa, the presence of ganglion cells but thinning (mild attenuation) of the muscle layers. Associated malformations seen in 7 (41%) of children were ambigious genitalia, vesicouretric reflux, absent left radias, pelvic kidney, bifid scrotum, hypospadias and a urogenital sinus. Postoperatively, there was no major complication except wound infection in a small number of infants. Although the pull-through procedure was followed by a degree of incontinence, the period of follow-up in most patients was too short to be assessed for the status and progress of continence.

DISCUSSION

The association of a high-type anorectal malformation with a pouch-like dilatation of the shortened colon was first described by Trusler et al in 1959.[8] Subsequently, a few sporadic cases were reported from time to time and given varied labels such as "absence of colon and rectum",[9] "cystic dilatation of colon",[10] "congenital atresia of anus with short colon malformation",[11] and "colonic reservoir".[12] Singh and Pathak presented six patients with short colon in 1972.[2] In 1984, Narsimaha Rao presented his series and proposed the name "Pouch Colon Syndrome" and suggested the anatomical classification of these patients into four different types.[6] In Kashmir, the first patient with pouch colon syndrome was recorded in 1984; a spurt in the number of cases

followed soon after. It is interesting to note that all our patients with high anorectal malformation were associated with pouch colon syndrome compared with a reported rate of 5% to 37%.[3,5,7,13,14] The anomaly is seen predominantly in the male and 76% of our patients had the type IV anomaly compared to other series in which type I and type II CPC constituted 80%.[14,15]

The marked regional variation in the incidence and the type is difficult to explain. It is possible that a racial (genetic), dietary or environmental factor, or a combination of these, is responsible.[2] The generally accepted theory of embryogenesis is defective positioning of the embryonic cloaca by a descending urorectal septum during fourth to sixth week of intrauterine life.[3,5,6,7,14,15,16,17] Variation in the timing of this defective organogenesis may be responsible for the length of normal colon proximal to the colonic pouch and the site of the fistula, with early arrest resulting in type I and type II CPC and later arrest in type III and type IV CPC. It is probable in some patients that the deletion of varying segment(s) of the normal colon is due to intrauterine obliteration of inferior and parts of superior mesenteric arteries. This has helped explain rare variants of double pouch, association with rectal atresia, and coexistence of segmental dilatation.[18]

The clinical presentation is like anorectal malformation in the early neonatal period with an absent anus and early abdominal distention caused by the distended colonic pouch. A wide fistulous communication of the pouch with the genitourinary system usually leads to meconuria or pneumaturia. However, typical radiological features on an invertogram or a plain radiograph of the abdomen are a single air-fluid level, occupying more than 50% of the width of abdomen.[13,14] In patients with a large size fistula, the bladder may produce an air cystogram.[13]

The management of this condition has been redefined; the early operation of a preliminary window colostomy of the pouch is historical and is no longer practiced because of certain complications. The choice of the primary procedure is based on the condition of the patient at the time of admission and the anatomy of the pouch. Tubular colorrhaphy is the preferred procedure in type I and type II CPC in order to preserve the absorptive area of the colon; this step is not necessary in the other two types where a reasonable length of colon is available. Definitive reconstructive surgery for these patients is best performed by the posterior sagittal route, with a mandatory additional abdominal component to accomplish the pull-through. The management protocol of these patients is shown in Figure 4. This approach has been broadly agreed on by all those dealing with CPC neonates. However, there is a compelling argument for single stage management of all pouch colon in newborns. Gangopadhyay et al,[19] in comparing their experience between staged management and primary single stage procedure, have shown significantly improved results for "continence and cosmesis, low morbidity and mortality, and considerably low cost". The neonates with delayed presentation are usually ill and there is appreciable early mortality. They may not be fit for an extensive one-stage primary surgery. We feel that the treatment choice should be individualized keeping in mind the concurrent urogenital and other abnormalities that would influence the surgical plan in these neonates.

The important objective of surgical management is fecal continence that improves with age,[19] hence long term follow-up must be emphasized.[17] In contradistinction to the outcome with high anorectal malformations, the incidence of fecal incontinence remains understandably high in these children because of anatomical characteristics such as the complete absence of normal distal bowel, shortening of the overall length of the intestines as well as the absence of an ileo-caecal valve (type I CPC). There is also the likelihood that a proportion of children will develop redilatation of the tubularized colon pouch in type I and II, after pull-through.[20] This is likely to add to the burden of overall management and attainment of a reasonable quality of life. Although treatable by resective, albeit-difficult surgery, it underscores the inherently abnormal development represented by a spectrum of neuromuscular abnormality of the pouch colon.[21]

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