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Congenital pouch colon in female subjects

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ABSTRACT

Over a period of 5-year (May 2000 to April 2005) 29 patients of congenital pouch colon (CPC) were managed by single pediatric surgeon in the Department of Pediatric Surgery of a tertiary hospital. Of these, 11 were girls (M: F: 1.6:1). Detailed anatomy could be studied in nine patients, were included in this study. Age of presentation in female subjects ranged 1 day to 8 years. On examination, eight of the nine patients had single perineal opening suggesting a very high incidence of association of cloaca in female subjects with CPC. Four had short urogenital sinus with colonic pouch opening in the posterior wall of urinary bladder. In other two patients, CPC terminated in short cloaca. Anomalies of mullerian structures such as uterus didelphus and septate vagina ware commonly encountered. Proximal diversion with or without pouch excision was done as the initial preliminary treatment for all those patients who presented in early life. Of the nine girls, two died after the preliminary surgery. Only five patients have undergone definitive surgery. Definitive surgery included abdomino-perineal pull-through of proximal normal colon (n=2), tubularization of pouch with abdomino-posterior sagittal- pull through (n=2) and abdomino-posterior sagittal urethra-vaginoanorectoplasty with pull through of tapered pouch colon in one patient. Two of these patients had concomitant bowel vaginoplasty. Three patients with tubularized pouches had constipation and spurious diarrhea. However, good pseudo-continence of bowel was achieved on regular bowel washes. The cosmetic appearance of perineum in all these three patients was acceptable. The patients in whom pouch excision was done had diarrhea and severe perineal excoriation resistant to conservative management.

KEY WORDS: Congenital pouch colon, cloaca, colorrhaphy

INTRODUCTION

Congenital pouch colon (CPC) is an unusual abnormality characterized by abnormal pouch-like dilatation of shortened colon associated with an ano-rectal malformation with or without a genitourinary fistulous opening. For unknown reasons maximum incidence of CPC is noted in Northern India. Though the entity is more frequent in males, CPC in females carries a special emphasis for its association with wide variety of complex genitourinary anomalies and also for the complexity of the surgical maneuvers required in the management. Limited literature is available on females with the pouch colon.[1-2] The largest review on the entity lacks exactitude in the categorization of these patients[1] and consequently, a clear-cut management protocol to be followed is not available. Frequently the entity remains unsuspected and is first diagnosed during the surgery. The lack of preliminary knowledge to the primary surgeons often ends up in additional surgical procedures, which complicates the further management.

The present study here is aimed at describing the various anatomical characteristics encountered in patients with female CPC. We also suggest a management protocol based on our own experience and the understanding from existing literature.

MATERIALS AND METHODS

A single pediatric surgeon in the Department of Pediatric Surgery of a tertiary hospital managed 29 patients of CPC over a period of 5-year (May 2000 to April 2005). Of these, 18 were males and 11 were females (M: F: 1.6:1). Of these 11 girls, patient-related characteristics were not completely known in two patients and they were excluded from study. A retrospective analysis of various patient-related characteristics with special reference to anatomical characteristics of internal and external genitalia, type of CPC (according to Narasimharao's[3] and

Wakhlu's[2] classification) and the level of poucho-genitourinary communication was done in the rest of the nine patients. The preliminary and the definitive surgical management along with their attendant outcome were also studied.

During the same study period, 19 female patients with cloaca were managed, of which 45% had CPC.

Age of presentation varied from 1 day to 8 years. Delayed presentations were seen in those who were decompressing well through the perineal opening.

External genitalia

On examination, eight of the nine patients had single perineal opening. One patient, who had blindly ending pouch, had two perineal openings with normally looking genitalia. Vestibule in all patients with cloaca was short and had receded appearance. In four of these patients, a very short common channel (approximately 0.5 cm long) leading to closely approximated orifices of hypospadiac urethra and vagina could be demonstrated on retraction of the margins of perineal opening [Figure 1]. Two patients had characteristic clover leaf-like appearance of cloacal opening. One patient had unusual fusion of labia majora posteriorly [Figure 2].

Type of pouch found at laparotomy

As per the classification given by Narsimha Rao *et al*.[3] Incidences of various types of pouches were:
Type I CPC - 4 patients
Type II CPC - 3 patients
Type III CPC - 2 patients.
According to Wakhlu's classification, only two patients had partial pouch (short) colon, i.e., normal colon proximal to pouch >8 cm in length.

Table 1: Internal genitalia anatomy noted in our patients

Internal genitalia anatomy	n
Uterus didelphus with septate vagina	6
Bicornuate uterus with normal vagina	1
Unicornuate uterus with L rudimentary horn	1
Normal	1

Level of termination of colonic pouch in genitourinary system

Of the nine patients, the level of termination of colonic pouch could be ascertained in seven patients. The colonic pouch was opening in posterior bladder wall in four patients. It terminated as short cloaca in two other patients. Colonic pouch ended blindly in one patient.

Internal genitalia

Anomalies of internal genitalia could be studied in all patients. In one patient, the internal genitalia were normal. [Figure 3] shows classical finding of uterine didelphus that was seen in six of our patients. The details of internal genitalia anomalies are shown in Table 1.

Correlation of our findings with Chadha's classification[1] of female CPC and Raffensperger's classification of cloaca[4] are shown in Table 2.

Management

Flow diagram depicting details of management of individual patients is shown in Figure 4. After preliminary surgery, only five patients have undergone definitive surgery and one patient is waiting for abdomino-posterior sagittal pull through. The patients with short urogenital sinus needed a cutback procedure to expose both the urethral and orifices [Figure 5].

RESULTS

One patient died of congenital heart disease after her preliminary surgery. Another patient died of unrelated cause one month after the surgery.

The follow-up period in our patients who had undergone definitive surgery range between 6 months and 4 years. Three patients with tubularized pouches had constipation

Table 2: Correlation of our findings with Chadha's classification[1] of female congenital pouch colon and Raffensperger's classification of cloaca[4]

Type of pouch (Narasimhan classification)	External genitalia	Site of fistula	Internal genitalia	Type of CPC (Chadha's classification)	Type of cloacal anomaly (Raffensperger' s Classification)
Type II	Short UG sinus	Colo-vesical fistula	Bicornuate uterus, single vagina	B	-
Type I	Short UG sinus	Colo-vesical fistula	Uterus didelphus with septate vagina	B	-
Type II	Short UG sinus	Colo-vesical fistula	Uterus didelphus with septate vagina	B	-
Type II	Short cloaca	Colo-cloacal fistula	Uterus didelphus with septate vagina	A	Raffensperger type 5
Type III	Short cloaca	Colo-cloacal fistula	Unicornuate uterus with L rudimentary horn	A	Raffensperger type 1c
Type I	Short UG sinus	Colo-vesical fistula	Uterus didelphus with septate vagina	B	-
Type I	Normal	Blind ending pouch	Normal	-	-
Type III	Common cloaca	Not known	Uterus didelphus with septate vagina	-	-
Type I	Common cloaca	Not known	Uterus didelphus with septate vagina	-	-

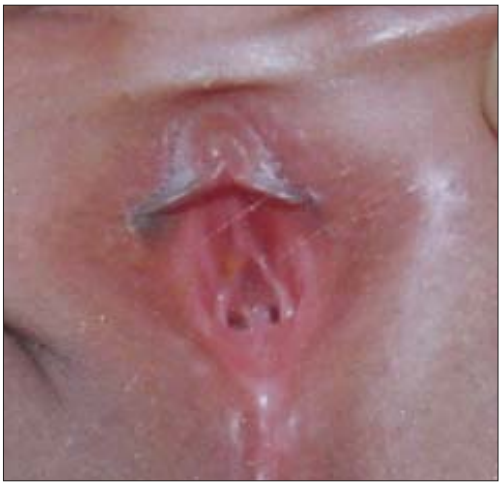

Figure 1: Ultra-short cloaca leading to urethral and vaginal openings

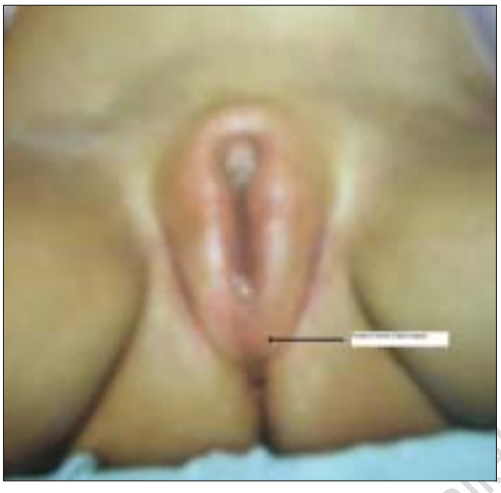

Figure 2: Posterior fusion of labia majora

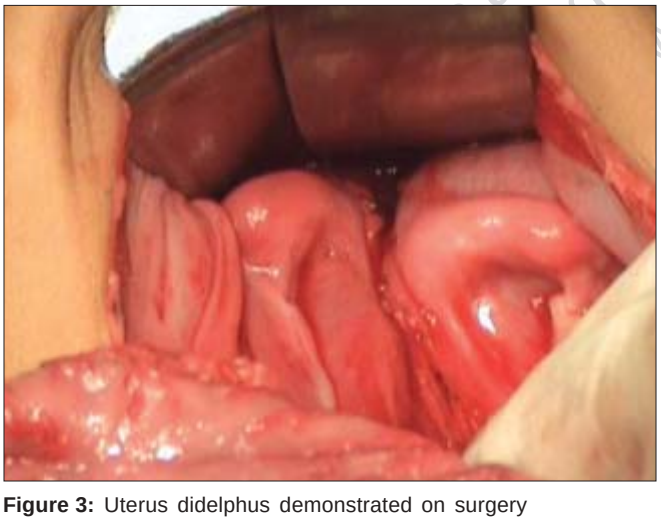

Figure 3: Uterus didelphus demonstrated on surgery

and spurious diarrhea. However, good pseudo-continence of bowel was achieved on regular bowel washes. The cosmetic appearance of perineum in all these three patients was acceptable. The eldest patient in this subgroup of patients is going to high school and is performing well. None of the patients had any metabolic problem. All of them have normal development for the age.

The patients in whom pouch excision was done had diarrhea and severe perineal excoriation resistant to conservative management [Figure 6].

DISCUSSION

Of the 360 reported cases of CPC in English literature, 26.67% have been known to occur in female subjects.[5] The relative incidence of CPC in female subjects in our study was 37.93%. Though complex, CPC in female has received less attention. This intricacies of this entity were addressed previously only by few authors.[1,2]

In our study, out of nine patients of CPC, eight had cloaca. Thus, the incidence of associated cloaca in female subjects with CPC was as high as 89%. The incidence quoted for concomitant presence of cloaca in female subjects with CPC in previously available literature ranges from 40 to 58%.[1,2,6]

The converse is also true. We have noted a 45% incidence of concomitant CPC in our patients presenting with cloaca. This suggests that at least in this part of the world, one should strongly suspect and rule out the presence of CPC in a case of cloaca. This is extremely relevant in view of the sitting a diverting colostomy in these patients. Similarly, if CPC is suspected on a plain roentgenogram in a female neonate with anorectal malformation, the possibility of one or the other subtype of cloaca should be kept in mind.

Of the eight patients with cloaca with CPC, one patient had type 5 cloaca (short cloaca) and another had type 1c cloaca (cloaca with septate vagina) (Raffensperger's classification).[4] In the other four patients, there was ultra-short UGS and the colon opened in the posterior wall of bladder through a fistulous opening. In two patients as the fistulous communication of bowel with genitor-urinary tract was not known, hence, we cannot classify them according to Raffensperger's classification.

Previous authors have described the fistulous communication in females with CPC as colo-claocal, colo-vaginal, colo-vesical and colo-vestibular.[1,2,6,7] But most of them have not mentioned the details of internal anatomy. Wakhlu *et al.* have casually mentioned cloaca and mullerian duct anomalies as separate entities in their 25 cases of CPC in female subjects.[2] Malformation of the mullerian ductal structures including all variety of duplex system, two separate uteri and vagina opening into a common channel or septate uterovaginal canal are

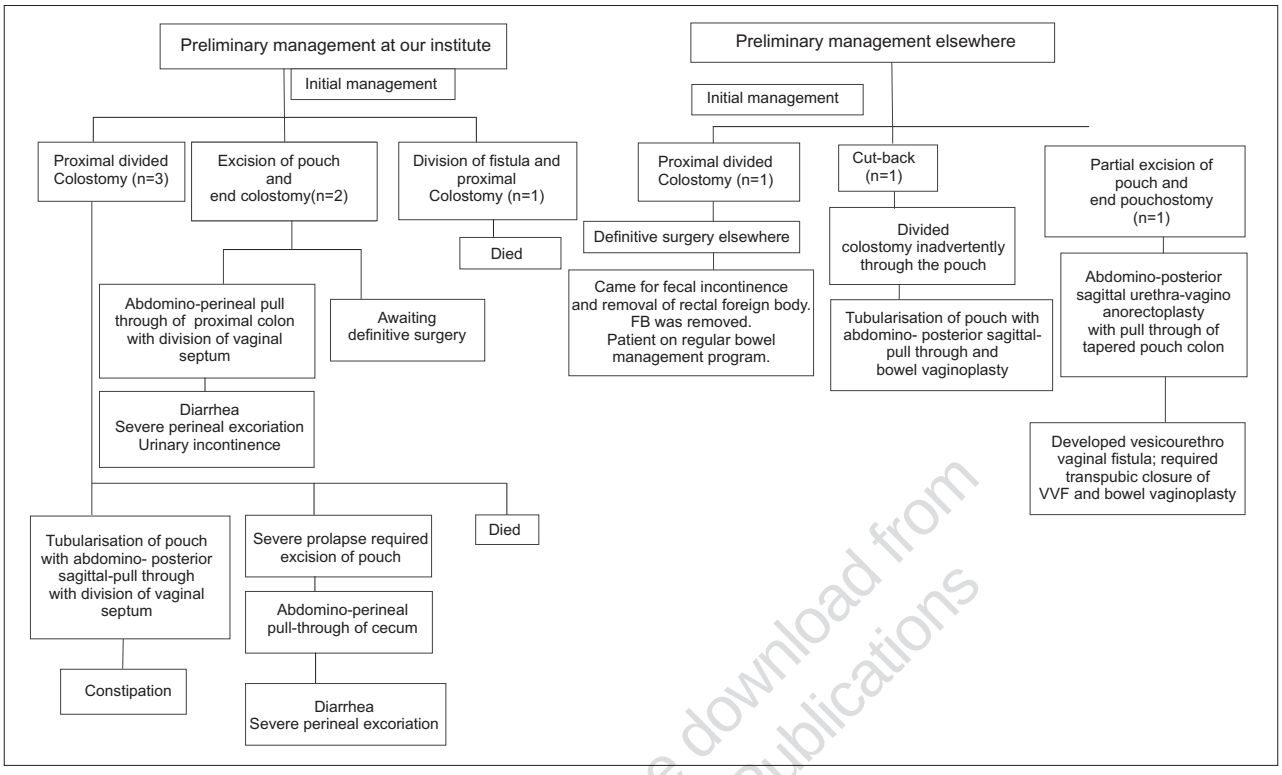

Figure 4: Flow diagram showing detailed management of nine patients

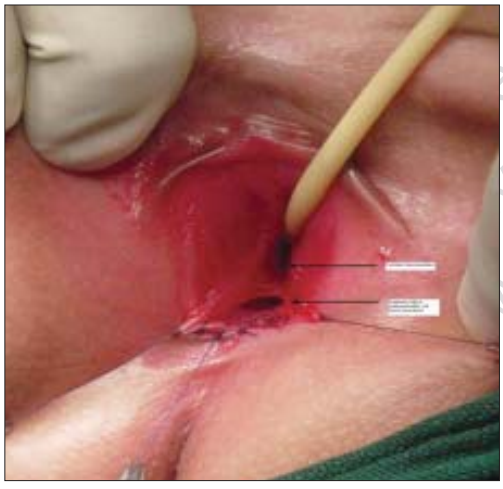

Figure 5: Cutback procedure done for short urogenital sinus

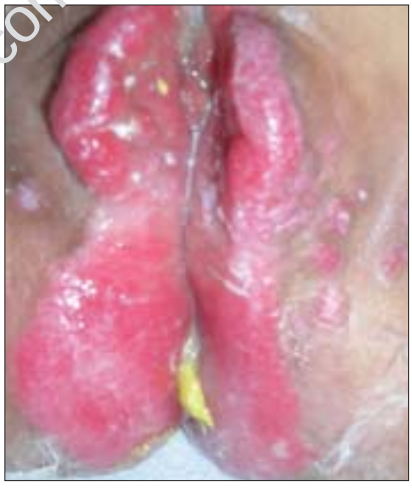

Figure 6: Severe perineal excoriation in patients where pouch excision has been performed

usual accompaniments of cloaca. So, it needs to be clarified whether the authors are talking about the same 10 patients of cloaca or different subset of patients. It is not surprising to note these discrepancies, as 'cloaca' is as such a neglected entity in this part of the world. A quick glance of search engines on internet would show gross paucity of literature about cloaca from India.

Chadha *et al.*[1] have detailed the various anatomical characteristics of CPC in female subjects and categorized these patients into three separate categories based on their level of fistula opening and type of CPC. The patients with CPC associated with persistent cloaca were grouped as Group A; Group B were the girls with Type I/II CPC with colo-vesical or colo-vestibular fistulas while type III/IV CPC with colo-vestibular fistulas were categorized as Group C. A thorough review of the literature revealed these descriptions to be in close semblance with the descriptions given for one or the other variant of cloaca, so there is nothing new described as far as type B is concerned. The description given in Chadha's category B was exactly similar to the description given for Raffensperger's cloaca type 9. The finding of colon opening into urethra-vaginal septum described as

characteristic feature seen only in females with CPC, has already been described by Hendren as one of the variant of cloacal anomaly years back.[8]

Chadha *et al.*[1] have quoted colo-vestibular fistula to be present in 60% subjects, but no patient in present series had colo-vestibular fistula. Most other authors have also quoted a far lower incidence of this entity (0-16%).[2,6,7] We tend to believe that Chadha's type C variant where the rectovestibular fistula has been described in association with type III and IV CPC, the anatomy in the vulva is that of an ultra- short cloaca or Raffensperger type 9 cloaca.[4]

We suspect that in most if not all of these cases of CPC in female subjects, one or the other type of cloaca is always present. In the light of this statement, we suggest a simplistic approach to treat it. Put most simplified, management of such patients is a management of two different anomalies- management of CPC and management of cloaca.

Treatment of cloaca, whether in single stage as proposed by Pena[9] or a staged management where initially the bowel is pull throughed and urogenital sinus is left untouched, is well described in English literature and any further discussion is beyond the scope of this manuscript.

With regard to management of CPC, majority believes in an initial 'minimalist' management of CPC in the form of proximal diversion. But then there is a large series published recently that recommends single-stage treatment of CPC.[10] Even if the traditional staged approach is followed, controversy prevails around preservation of colon and subsequent colorrhaphy. Colorrhaphy was first described by Trussler,[11] and it was appreciated and adopted by many in 1990s. But last few years have seen the upsurge of an opposite view as few authors have reported re-dilatation of tubularized pouch.[12,13] There is a mention about disorganized muscular arrangement that causes hypoperistalsis.[14] Gangopadhyay *et al.* has recommended colorraphy of only 5 cm of distal pouch in cases of complete pouch colon.[10] On the other hand, pouch excision in our experience has resulted in intractable diarrhea and severe perineal excoriation [Figure 6]. This issue is important for male subjects too but in females, excoriation results in severe vulvitis, vaginitis, and urethritis with its attendant morbidity. We believe that management of constipation with regular bowel washes is much easier than the management of severe excoriation and diarrhea. Rattan *et al* has recently mentioned a technique of 'pouch colon patch graft'[15] that seems worthy of a trial.

As individual experience from different centers is miniscule, there is urgent need to have a national registry about the entity. Only collective efforts from many centers can make us wiser about the embryology, anatomical details and management of this unique entity. Our hypothesis based on our small experience that the cloaca is present in most cases of female subjects of CPC needs to put to be test.

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