Original Article

Congenital pouch colon in girls: Genitourinary abnormalities and their management

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ABSTRACT

Aims: To discuss the assessment and management of genitourinary (GU) tract abnormalities in 21 girls with Types I-III congenital pouch colon (CPC), studied over a period of 10 years. **Materials and Methods:** Assessment included clinical and radiological assessment, examination under anesthesia (EUA), endoscopy of the lower GU tract, and evaluation of the surgical findings, operative procedures for the GU anomalies, and the results of management. **Results:** Initial examination of the external genitalia showed a "clover-leaf" appearance (n = 6) and a single perineal opening (n = 6). In 9 patients, the openings of the urethra and double vagina were seen, of which a vestibular fistula was seen in 5 and an anterior perineal fistula in 1. Seventeen patients (81%) had urinary incontinence (UI) - partial in 10, and complete in 7. Renal function tests, X-ray sacrum, and abdominal US were normal in all patients. Micturating cystourethrogram (n = 9) showed a wide, bladder neck incompetence (BNI) with reduced bladder capacity in seven patients. EUA and endoscopy revealed a septate vagina in all patients and the urethral opening at a "high" position (n = 14) or at a relatively normal or "low" position (n = 7). In 8 patients, the intervaginal septum was thick and fleshy. Endoscopy showed a short, wide urethra, an open incompetent bladder neck, poorly developed trigone, and reduced bladder capacity in the patients with UI. The fistula from the colonic pouch opened in the proximal urethra (n = 4), high in the vestibule (n = 3), low in the vestibule (n = 8), perineum just posterior to the vestibule (n = 1), and undetermined (n = 5). Vaginoscopy (n = 8) showed normal cervices in all and cervical mucus in 4 patients. The subtypes of CPC were Type I CPC (n = 4), Type II CPC (n = 16), and Type III CPC (n = 1). All 21 patients had uterus didelphys. In four patients with UI, during tubular colorraphy, a segment of the colonic pouch was preserved for later bladder augmentation if required. A Young-Dees bladder-neck repair (BNR) was performed in four older girls for treatment of UI, with marked improvement in urinary continence in two girls, some improvement in one girl with complete urinary incontinence, and minimal improvement in one child. Division of the intervaginal septum was performed in three girls. **Conclusions:** GU abnormalities in girls with CPC need to be assessed and managed by a tailored protocol. UI is frequent, and its correction may require BNR. A segment of the colonic pouch can be preserved for possible future bladder augmentation. All girls have a septate vagina, often widely separated, and uterus didelphys. Gynecologic assessment and monitoring is required throughout adult life. Considering the wide opening of the vestibule, surgical management of the urogenital component by division of the intervaginal

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septum and if required, the vagino-fistula septum on each side results in a normal or a hypospadiac urethral opening and an adequate vaginal inlet.

KEY WORDS: Anorectal malformation, congenital pouch colon, urinary incontinence, uterus didelphys, septate vagina

INTRODUCTION

Congenital pouch colon (CPC)[1] is an unusual abnormality in which a pouch-like dilatation of a shortened colon is associated with an anorectal malformation (ARM). CPC has most frequently been reported from centers in northern India[1-7] where it comprises 4.38%[2] to 18.7%[7] of all ARM. The condition is less common in females, the male to female ratio in large series ranging from 2.25:1[5] to 7:1.[6] The anomalous anatomy in girls with CPC has not been described in detail in most series reported in the literature. An earlier study from our center[8] found that girls with Types I-III CPC had a septate or double vagina, uterus didelphys, and a high incidence of urinary incontinence (UI) due to an open or bladder neck incompetence (BNI) and a short urethra. The colonic pouch opened by a fistula either into the urethra or in the vestibule in a "low" or a "high" position.[8] The present follow-up study details the assessment and management of genitourinary (GU) abnormalities identified in 21 girls with Types I-III CPC over an 11-year period. The discussion centers on the management of specific GU abnormalities.

MATERIALS AND METHODS

During the period from January 2003 to March 2014, 41 girls with CPC were managed by the Department of Pediatric Surgery, Lady Hardinge Medical College and Kalawati Saran Children's Hospital, New Delhi. 20 of these 41 patients presented during the neonatal period or infancy underwent preliminary assessment and management but were then either lost to follow-up or are as yet awaiting detailed assessment.

The remaining 21 patients were evaluated in more detail regarding the status of the GU tract. Assessment included clinical and radiological assessment, examination of the external genitalia and perineum examination under anesthesia (EUA), endoscopic examination of the lower GU tract, and evaluation of the operative findings and procedures performed. This group includes 12 patients who were part of the study material in an earlier report from our center[8] and had remained on follow-up. The remaining nine patients had presented after the previous study had been completed. The age ranges of the 21 patients at assessment as well as the main parameters assessed are shown in Tables 1 and 2.

The protocol for radiological assessment consisted of X-rays of the sacrum, an abdominal ultrasound (US), and a micturating cystourethrogram (MCU). However,

Table 1: Urogenital abnormalities in girls with CPC: Clinical features, and findings on EUA and endoscopy (*n* = 21)

Parameter	*n* (%)
Age at assessment	Range 6 months-14 years (mean 4.03 years)
External genitalia (outpatient examination)	
"Clover-leaf" appearance	6
Ur., Sept. V	3
Ur., Sept. V, VF	5
Ur., Sept. V, anterior perineal fistula	1
SPO	6
Urinary continence	
Normal	4
PUI	10
CUI	7
EUA: Septate/double vagina	21 (100)
EUA-position urethral opening	
"High" (>1.5 cm from base of clitoris)	14 (in 6, 'very high')
"Low" (<1.5 cm from base of clitoris)	7
EUA/endoscopy-location of fistula (*n*=19)	
Not seen	5
APF	1
Low VF	8
High VF	3
Proximal urethra	4
Operative findings	
Double uterus, vagina (uterus didelphys)	21 (100)

SPO: Single perineal opening, Ur., Sept. V: Urethral opening with septate vagina, VF: Vestibular fistula, PUI: Partial urinary incontinence, CUI: Complete urinary incontinence, EUA: Examination under anesthesia, CPC: Congenital pouch colon, APF: Arteriopelvic fistula

Table 2: Correlation of cystourethroscopy findings of bladder neck and urethral length with urinary continence (*n* = 19)

Bladder neck	Urinary continence	Urethral length
Normal (*n*=2)	Normal (*n*=2)	Short (*n*=2)
BNI (*n*=17)	CUI (*n*=7)	Short urethra (*n*=2)
		Very short urethra (<1 cm) (*n*=5)
	PUI (*n*=10)	Short urethra (*n*=5)
		Very short urethra (*n*=5)

BNI: Bladder neck incompetence, CUI: Complete urinary incontinence, PUI: Partial urinary incontinence

due to logistical reasons, a MCU could be attempted in only nine patients, including six patients from the previously reported series,[8] largely because in several patients the urethral opening was not clearly identifiable. In five patients, an intravenous urogram (IVU) had been performed elsewhere, prior to referral to our center.

At surgery, both during the preliminary and subsequent surgeries, the anatomy of the pouch colon malformation, the external and internal genitalia, and the urinary tract were recorded. On the basis of operative findings, patients were categorized into the four subtypes of CPC described by Narasimharao *et al*.[3] The operative findings and procedure(s) performed as well as the results of management were recorded in detail, especially with regard to the associated GU abnormalities.

RESULTS

The results of the study are summarized in Tables 1-3 and detailed below.

CLINICAL FINDINGS

External genitalia

In all 21 patients, on outpatient examination, without retracting the labia, the external genitalia and perineum appeared essentially normal without any of the features characteristic of cloacal malformations [Figure 1]. In 9/21 patients (42.9%), the posterior margin of the vestibule appeared wide [Figure 1].

On retracting the labial folds, a "clover-leaf" appearance,[9] described in detail in an earlier report,[8] was seen in six patients [Figure 2a]. In nine patients, the openings of the urethra and the septate vagina could be visualized [Figure 3]. In 5 of these 9 patients, the opening of a vestibular fistula (VF) could also be identified [Figure 4] while one girl had an anterior perineal fistula [Figure 5]. In six patients, only a single, but wide, single perineal opening (SPO) was seen [Figures 6a and 7a].

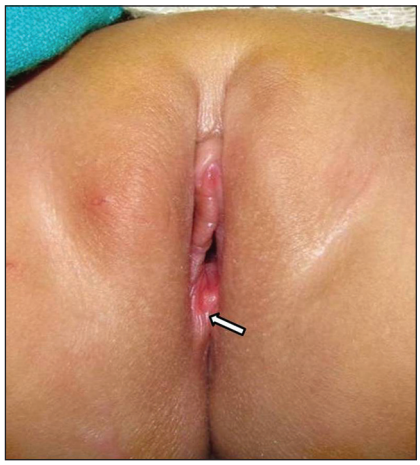

Figure 1: Normal appearance of the external genitalia in a girl with congenital pouch colon. Note the wide posterior margin of the vestibule (arrow)

Urinary continence

Totally, 17 of the 21 patients (81%) had UI which was partial in 10 girls' partial urinary incontinence (PUI) and complete urinary incontinence or near-complete (CUI) in seven others. The girls with PUI were passing urine in a stream at regular intervals but also had dribbling of urine. Extensive perineal and perivulval excoriations due to UI were present in five of the seven girls with CUI and in two girls with PUI [Figure 6a]. Three patients were thought to be the continent of urine at initial presentation during infancy but during later assessment at age ranging from 2 years to 3 years, CUI was apparent in two girls and PUI in one girl. In two patients, the parents felt that the degree of UI had worsened with increasing age of the child.

BIOCHEMICAL AND RADIOLOGICAL INVESTIGATIONS

Biochemical renal function tests were normal in all 21 patients. X-rays of the sacrum were normal in the 20 patients in whom films were available for assessment. Abdominal US showed normal kidneys and ureters in all patients. MCU, attempted in 9 patients, was unsuccessful

Table 3: Operative procedures for GU anomalies

Procedure	*n* (%)
Low ligation fistula from colonic pouch (without dissection from trigone/urethra)	16 (72.9)
Preservation of segment of colonic pouch during TC (for possible bladder augmentation) in girls with UI	4
Young-Dees BNR for UI	4
Division of intervaginal septum	3 (14.3)

TC: Tubular colorraphy, BNR: Bladder-neck repair, UI: Urinary incontinence, GU: Genitourinary

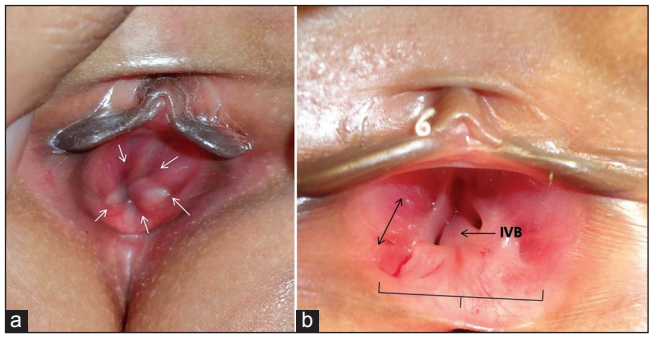

Figure 2: Outpatient department examination showing "clover-leaf" appearance of the external genitalia (arrows), and (b) examination under anesthesia showing a "very high" urethral opening, double vagina, wide inter-vaginal bridge, tilting of vaginas (double-headed arrows), and wide posterior margin of the vestibule (bracket). The fistula from the colonic pouch opened in the proximal urethra

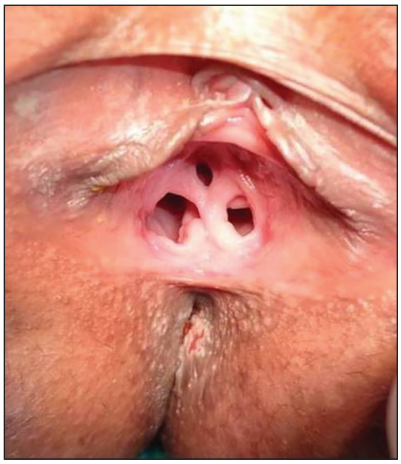

Figure 3: Outpatient department examination showing a "low," wide urethral opening, and septate vagina; fistula not identified on endoscopy

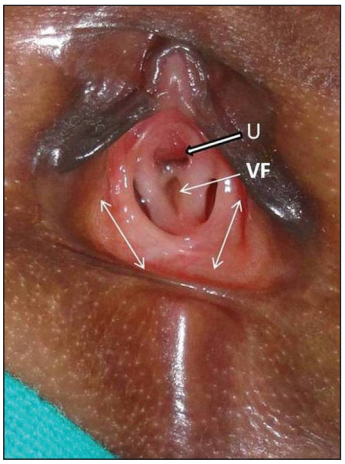

Figure 4: Outpatient department examination showing a "low," wide urethral opening (U), low anteriorly placed vestibular fistula, double vagina (double-headed arrows show tilt of vaginas)

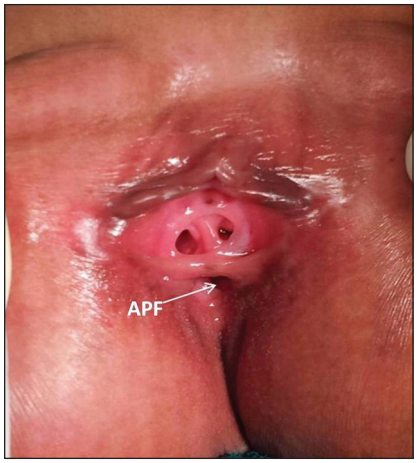

Figure 5: Outpatient department examination showing urethra, double vagina, and an anterior perineal fistula

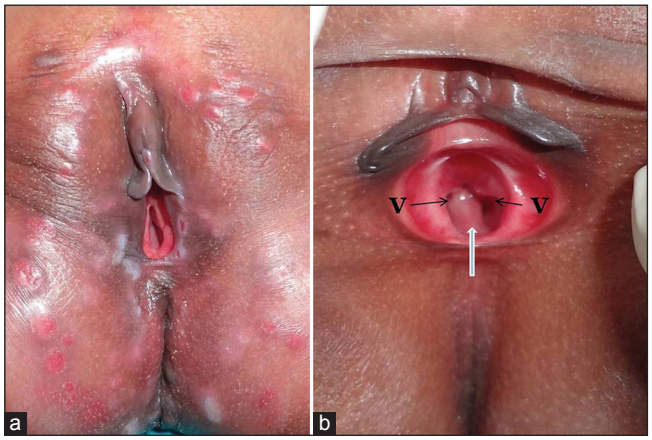

Figure 6: (a) Outpatient department examination showing a single perineal opening with perivulval excoriations due to partial urinary incontinence, and (b) examination under anesthesia showing a "very high" urethral opening with a "high" vestibular fistula, double vagina (V) and a wide intervaginal bridge (arrow)

in one patient with CUI as the dye could not be retained. An open bladder neck and decreased bladder capacity was evident in seven cases, five of whom had PUI while two had CUI. In three of the seven girls, the urethra was short and wide [Figure 8]. In another girl with PUI, the MCU showed good bladder capacity with a less widely open bladder neck. In three patients, the MCU showed bilateral Grade III vesicoureteric reflux (VUR) in two girls and Grade II VUR in one girl [Figure 8]. In one girl with Grade III VUR, both ureters were seen to open low down, close to the bladder neck. In one girl, the MCU showed filling up of the two vaginas. IVU ($n = 5$) showed a unilateral bifid collecting system in two patients.

EXAMINATION UNDER ANESTHESIA AND CYSTOURETHROGENITOSCOPY

Urethral and vaginal openings

Examination under anesthesia and endoscopic examination confirmed a septate/double vagina in all 21 patients. This included the patients in whom only a "clover leaf" appearance ($n = 6$) [Figures 2b and 9] or an SPO ($n = 6$) [Figures 6b and 7b] had been seen on outpatient examination. The urethral opening and thus, the meeting point or confluence of the urethral and vaginal openings, was classified as being "low" or near-normal (<1.5 cm from the base of the clitoris) [Figures 3 and 4] or "high" (>1.5 cm from the base of the clitoris) [Table 1].[8] In 6 patients, the urethral opening was "very high"/severely hypospadic [Figures 6b and 7b]. In one child, the urethral meatus could only be seen on wide retraction of the margins of the vulval vestibule [Figure 10]. On EUA, the urethral meatus was wider than normal in all patients [Figures 3 and 4]. In some cases, the vaginal openings were tilted with the posterior ends diverging inferiorly and laterally [Figure 2b and 7b] while in some other patients, the openings were tilted in the opposite direction with the anterior ends diverging laterally and

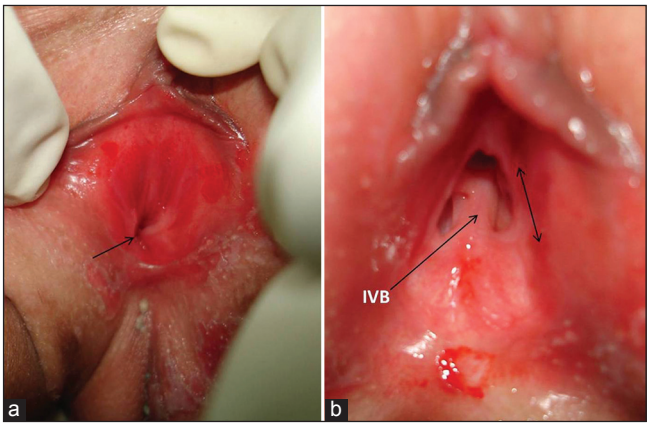

Figure 7: (a) Outpatient department examination showing a single perineal opening (arrow), and (b) examination under anesthesia showing a "very high" urethral opening, double vagina, and a wide inter-vaginal bridge. The fistula was not seen on endoscopy. Double-headed arrow shows tilt of vaginas

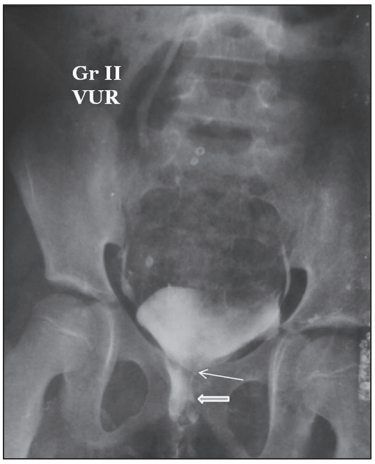

Figure 8: Micturating cystourethrogram showing a small capacity bladder, bladder neck incompetence (arrow), short, wide urethra (wide arrow), and bilateral Grade I/II vesicoureteric reflux

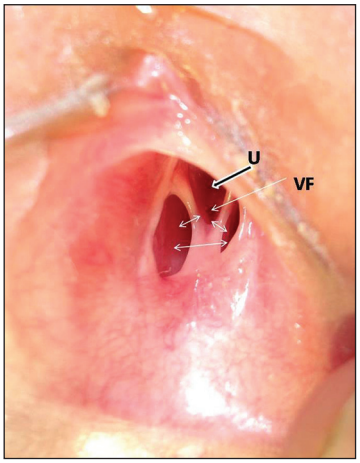

Figure 9: Examination under anesthesia showing a "high" wide urethral opening (U), a narrow "high" vestibular fistula/distal urethral fistula, and double vagina. Double-headed arrows show line of division of intervaginal bridge and vagino-fistula septum on each side

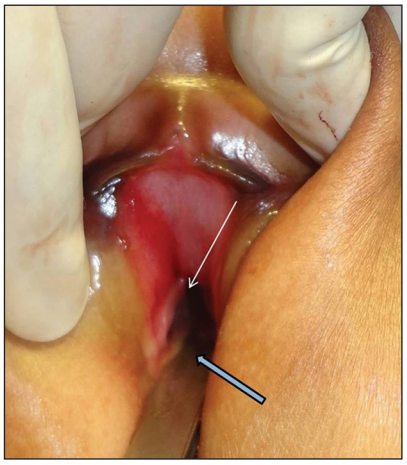

Figure 10: Examination under anesthesia showing a "very high" urethral opening (arrow) visualized only after insertion of retractor in the wide introitus (wide arrow)

the posterior ends meeting in the midline [Figure 4]. In two girls, the two vaginas were close together, without any divergence, and the VF or anterior perineal fistula was located just posterior to the margin of the vaginas [Figure 5]. The intervaginal septum was wide and thick in eight patients [Figures 3, 4, 6b, and 7b].

The position of the urethral opening did not appear to correlate with urinary continence. Of the six patients with a "very high" urethral opening, two (50%) had normal urinary continence, two had PUI, and only one child had CUI. Of the seven patients with a "low" urethral opening, three had CUI, three had PUI, and only one had normal continence.

Endoscopic findings

Endoscopic examination (n = 19) showed that the urethra was short in length (usually <2 cm long) in all patients including two with a normal bladder neck and normal urinary continence [Table 2]. In 10 patients, the urethra was very short, <1 cm in length. All these patients had UI and included five of the seven patients with CUI and 5 of the 10 patients with PUI. An open or BNI was present in 17 patients with UI [Figure 11]. Table 2 summarizes the correlation between the endoscopic findings of the bladder neck and urethral length with urinary continence. In several patients, the short urethra and the bladder-neck appeared to be deficient posteriorly [Figure 12]. In another patient, a midline ridge was visible on the posterior wall of the bladder probably representing the closely apposed terminal fistula of the colonic pouch.

In addition to BNI, the trigonal area was poorly developed, and the ureteric orifices were clearly identified in only six patients. In all six patients, the ureters were opening laterally and low down, close to

the bladder neck [Figure 12]. Although the assessment was subjective, bladder capacity was reduced in 16 patients and appeared markedly reduced in four of these 16 patients.

Vaginoscopy (n = 8) showed normal appearing cervices in all patients with cervical mucus visible in four patients. Two of the older patients had had a normal puberty and were having normal menstrual periods.

Terminal fistula of the colonic pouch

On EUA and/or endoscopy, the fistulous opening of the colonic pouch could not be identified in five patients, in all of whom the fistula had been ligated during an earlier surgery. In four patients, the fistula opened into the proximal urethra just distal to the bladder neck. In the remaining 12 patients, the opening of the fistula was just posterior and a little proximal or cranial to the urethral opening, anterior to the vaginal orifices in three patients [Figure 9], a position that can be described as "high vestibular" or "distal urethral." Eight patients had a "low VF" [Figure 4], while one patient had an anterior perineal fistula [Figure 5]. Endoscopic examination of the VF in four patients in whom the colonic pouch had been ligated during preliminary surgery showed it to end 2-5 cm proximally.

OPERATIVE FINDINGS

The subtypes of CPC were Type I (n = 4; 19%), Type II (n = 16; 76.2%), and Type III (n = 1; 4.8%). All 21 patients (100%) had 2 hemi uteri with one hemi uterus and vagina flanking the termination of the colonic pouch on each side [Figure 13]. Varying degrees of distension of the colonic pouch were noted. An unusual finding was that in 7 patients, one with Type I CPC and 6 with Type II CPC, the urinary bladder appeared large, lobulated and more flaccid than normal [Figure 13].

GENITOURINARY TRACT-OPERATIVE PROCEDURES PERFORMED

In 16 patients, during preliminary/definitive surgery, the fistulous termination of the colonic pouch was ligated as low down in the pelvis as comfortably possible [Table 3]. A proximal ileostomy was the preliminary surgery in seven girls. Definitive surgery for management of the CPC consisted of pull-through of the tubularized colonic pouch (n = 9), pull-through of normal ileum/colon proximal to the pouch (n = 4), while the parents of two patients, one with an end colostomy after tubularizing colorraphy (TC) for a Type II CPC and another with an end-colostomy after excision of a Type II pouch, opted for a permanent abdominal end-colostomy.

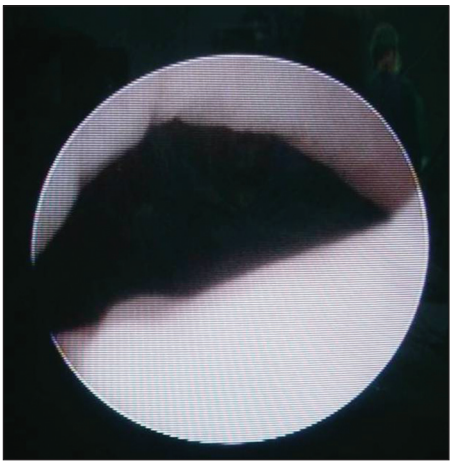

Figure 11: Cystourethroscopy showing a widely open bladder neck incompetence in a girl with urinary incontinence

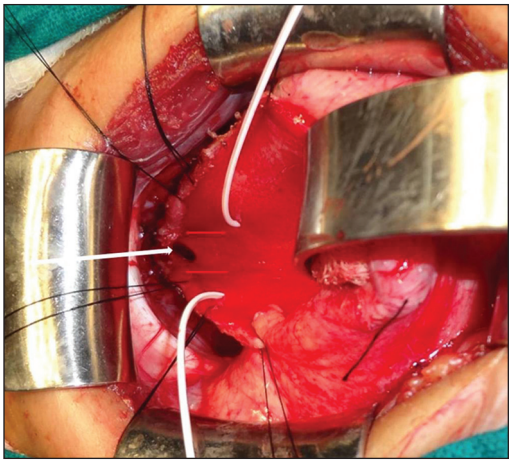

Figure 12: Operative photograph during bladder-neck repair showing widely open bladder neck, deficient posteriorly (arrow), and laterally placed ureteric openings. Lines mark incisions for tubularization of trigone

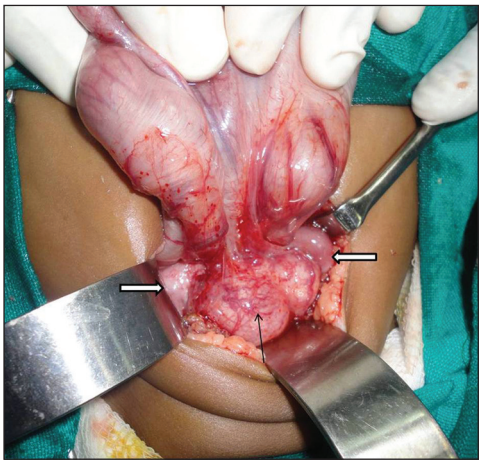

Figure 13: Operative photograph showing the colonic pouch, double uterus with distended vaginas (broad arrows), and a flaccid lobulated urinary bladder (fine arrow)

Four patients were lost to follow-up while one child is awaiting definitive surgery.

During the course of this study, it became apparent that the vast majority (81%) patients had either PUI or CUI. In four patients, managed during the latter part of the study period, a segment of the colonic pouch was retained so that, if required, it could be available for bladder augmentation to treat UI. In three of these patients, during definitive surgery, the fistula was ligated low down in the pelvis. The lateral or outer portion of the colonic pouch was tubularized (TC) and while preserving its leash of vessels, the remaining portion of the pouch was retained [Figure 14]. An abdominal stoma was created for this otherwise blindly ending pouch segment while pull-through of the tubularized colon was performed. In a 5-year-old girl with Type II CPC and PUI, pull-through was performed after TC, leaving a segment of the colonic pouch *in situ*, communicating with the VF. These four patients have as yet not undergone any definitive surgery for the UI.

Definitive surgery for CUI has been performed in 4 girls, the age at surgery ranging from 5 to 12 years [Table 3]. In all patients, surgery revealed a wide bladder neck and laterally placed ureteric openings. In an 8-year-old girl, the bladder neck was found to be widely open with a deficient posterior margin of the urethra and bladder neck, the urethral opening being visible [Figure 12]. A Young-Dees bladder-neck repair (BNR) was performed over a 10 Fr catheter [Figure 12] along with, in one girl, reimplantation of the ureters at a more proximal level to gain more length of the bladder neck and trigonal area for the bladder neck tightening. Postoperatively, there is marked improvement in urinary continence with diurnal dry intervals ranging from 3 h to 4 h in 2 girls, improvement from CUI to a diurnal dry interval of 1-2 h in one girl, and no/minimal improvement in one child.

Division of the intervaginal septum has been performed in 3 girls during definitive pull-through surgery. In one girl, this necessitated intervaginal anastomosis for mucosal approximation. In another girl, division of the vagino-fistula septum on each side was also performed.

One patient who had undergone division-ligation of the "high" VF developed a vesical calculus extending in a dumbbell fashion through the bladder neck into the distal retained remnant of the colonic pouch. The stone as well as its extension were removed by a suprapubic approach.

DISCUSSION

The commonly seen anomalous anatomy of the GUT and the terminal fistula of the colonic pouch are shown in Figure 15.

Although similar anatomic findings have been described earlier,[2,8-16] none of these studies have reported endoscopic findings regarding the urethra, integrity of the bladder neck or the fistula of the colonic pouch.

It is interesting that cases of persistent cloaca with a very long common channel have been described where the rectum opens into the bladder neck or trigone, just anterior to the openings of two completely separated vaginas, one on each side of the high rectal fistula.[17-19] Apart from a long, narrow cloacal channel, this anatomy is strikingly similar to that described here. There is also a similarity with the anatomy in cases of cloacal exstrophy in girls[20] and a marked similarity with that in patients of "completely covered cloacal exstrophy."[21,22]

The management of GU abnormalities, discussed in the following section, requires special consideration especially as no management protocol has been described earlier. It is important during initial examination to suspect the anomaly on the basis of the

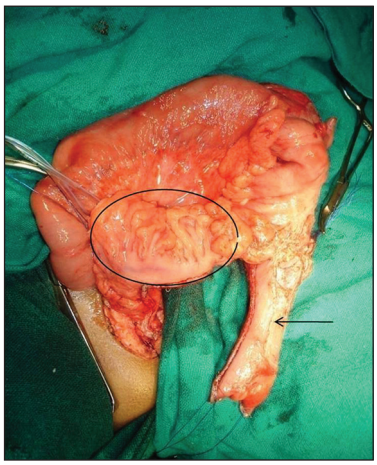

Figure 14: Preservation of a segment of the colonic pouch for possible future bladder augmentation in a girl with urinary incontinence (ellipse). Arrow shows the tubularized colon for pull-through

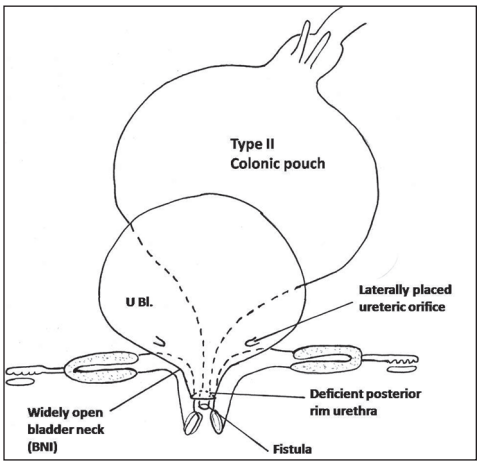

Figure 15: Diagrammatic representation of the usual anatomy in a girl with Type II congenital pouch colon

characteristic anatomy of the external genitalia.[8] UI may also be present. Abdominal X-rays usually show a normal sacrum and the large air-fluid level characteristic of CPC.[1,3,5-8] In older girls, a large soft-tissue shadow representing fecal matter inspissated in the distended colonic pouch may be present. In patients with a VF in whom CPC is suspected, a contrast enema can show the anatomy of the colonic pouch.

MANAGEMENT OF THE COLONIC POUCH AND ITS TERMINAL FISTULA IN RELATION TO THE GENITOURINARY ABNORMALITIES

The terminal portion of the colonic pouch and its fistula is usually closely apposed to the posterior wall of the urinary bladder in the trigonal area and the urethra. Thus, a complete dissection of the fistula from these structures carries a high risk of damage to the trigone and urethra, and their nerve supply. The fistula should be ligated as low-down in the pelvis as is safely possible. The distal remnant of the fistula remains as a blind-ending tract opening into the urethra or the vestibule.[8,10,14] Low ligation of the fistula is important as one of our patients presented with a stone extending from the colonic pouch remnant into the bladder through the high VF.

Several surgical options have been described for the initial management of the colonic pouch.[1,3-5] In our series, several patients underwent a preliminary ileostomy which is a good option in newborns and infants in whom it may be difficult to assess urinary continence as well as the detailed anatomy of the GU tract. At a later stage, GU assessment can help decide whether a portion of the colonic pouch needs to be preserved for possible future bladder augmentation as part of treatment for UI. Constructing an ileostomy, however, necessitates the management of ileostomy losses for prolonged periods with problems of dehydration, electrolyte and metabolic disorders, nutritional deficiencies, and peristomal excoriations.[23,24] This is especially a problem in developing countries like India. In Type III CPC, a proximal colostomy may, therefore, be a better option. In girls presenting beyond infancy, detailed assessment and a definitive management protocol can be implemented at the presentation itself.

MANAGEMENT OF ABNORMALITIES OF THE GENITOURINARY TRACT (GENITOURINARY COMPONENT OF THE ANOMALY)

Prior to definitive management, it is important to perform an abdominal US, an MCU, and EUA with an endoscopic examination of the lower GU tract and the fistulous opening. MCU may be difficult to perform in patients with a "high"/"very high" urethral opening which is difficult to identify and also if there is severe BNI with gross leakage of the contrast.

Renal and upper tract abnormalities

Significant upper urinary tract abnormalities are uncommon. Although VUR has been reported to be very common in patients of CPC,[1,3,5] MCU showed significant bilateral VUR in only 3/9 girls. However, it is difficult to know the exact incidence as often, a severely deficient bladder neck may result in an incomplete study. Although upper tract abnormalities are not frequent, an IVU or MR Urogram would help provide a more complete assessment.

Management of urinary incontinence

The anatomical features responsible for UI are a partially or widely open bladder neck (BNI) which may be deficient posteriorly [Figure 12], and reduced bladder capacity. The short, wide urethra may also contribute to UI. Due to the frequent "high" confluence of the urethral and vaginal openings and the tilt of the vaginas, vaginal pooling of urine after micturition is common and can worsen UI.

The degree of UI varies greatly and may worsen with age. However, the level of the urethral opening does not always correlate with UI or its severity. The integrity of the bladder neck, even in the presence of a short urethra, appears to be the main factor deciding urinary continence.[8] This is shown by the two patients with normal urinary continence who had a short urethra with a normal bladder neck. Although all the 10 patients with a very short urethra (<1 cm) had UI, this did not correlate with the severity of UI as these included 5 patients with PUI as well as 5 girls with CUI [Table 2].

There are no earlier reports regarding the management of UI in these patients. An exception is a girl with Type I CPC, double uterus and vagina, a perineal fistula, and CUI thought to be due to bladder hypoplasia in whom UI was managed by reimplantation of the ureters into an ileal conduit.[13]

In our series, a Young-Dees BNR was performed in four patients, with good results in 2 girls and some improvement in one child. Reimplantation of the lateral and low-placed ureters at a higher level was necessary to gain more length for tubularization of the trigone in only one girl, underlining the anatomical variations that may be present. However, the bladder capacity and anatomy were largely suitable for BNR in all girls. Significantly, unlike patients with persistent cloaca in whom splitting of the pubic symphysis may be required to gain access to the bladder neck and urethra for BNR,[17,25] in our

patients, this was not necessary because of the "high" urethral opening and the short urethra. Apart from this technique, other procedures to augment bladder-outlet resistance may need to be considered as alternative or additional procedures.

It is possible that some patients undergoing BNR, especially those with small-capacity bladders may require bladder augmentation to achieve satisfactory urinary continence. In Types I-II CPC, normal proximal colon is either absent (Type I CPC) or very short (Type II CPC), and a segment of the colonic pouch is usually preserved by TC for increasing the colonic absorptive area.[1,4-6] Although we have no personal experience, if required, bladder augmentation may necessitate the use of the stomach or small bowel as described for patients with CE.[23,24] One useful feature in Types I-III CPC is that as in four of our patients with UI, the portion of the colonic pouch remaining after TC can be preserved for later bladder augmentation.

Importantly, a high and inaccessible urethral opening may make clean intermittent catheterization (CIC) difficult or impossible in several patients. CIC would be mandatory after bladder augmentation and at times even after simple BNR. It may also not be possible to use the appendix as a catheterizable channel by a Mitrofanoff procedure[26] as it is often absent/short and stubby or else duplicated.[1,3-5] In addition, the anatomy of the pouch colon and its surgical management may preclude the use of the appendix. A Monti procedure[27] may, therefore, be necessary for creating a catheterizable channel. If procedures to provide effective bladder outlet resistance with or without bladder augmentation do not achieve satisfactory results, an option may be to surgically close the bladder neck and create a catheterizable channel for emptying the bladder or reservoir.

Management of the urogenital sinus anomaly and septate/double vagina

Management for the urogenital (UG) sinus anomaly and abnormalities of the genital organs has not been described in detail in reports from India. Most centers have described cloacal reconstruction[1,4,5,11] without details of the anomalous anatomy or the procedures performed. A recent report[28] described managing girls with CPC and a cloaca either by PSARVUP or by "total UG mobilization."[29] Another report also described total UG mobilization for the management of the UG anomaly.[30] At our center, earlier these cases were misdiagnosed as having "persistent cloaca." In 4 patients, surgery consisted of tubularization of the anterior wall of the common channel to extend the hypospadiac urethra to the perineum and a small bowel graft to extend the two vaginas to the perineum.[1,11] This extensive surgery is tedious, difficult, has a high risk of complications and as shown by our findings, is unnecessary. The only likely benefit is that extending the urethra further down to the perineum may make the urethral meatus more accessible for catheterization and reduce vaginal pooling of urine.

Perhaps the most important finding of our study is that these patients do not have a "persistent cloaca" in the accepted sense of the term. Surgery for the UG anomaly is not required apart from division of the intervaginal septum and if feasible (in cases with a "low" VF), the vagino-fistula septum on each side as described by Demirogullari *et al*.[14] [Figures 4 and 9]. This, considering the wide vestibule, will leave the child with an adequate vaginal opening and a wide vagina. In girls with a "low" urethral opening, the urethral meatus is near-normal in location. Girls with a "high" urethral opening will be left with a hypospadiac meatus [Figure 10] which should be acceptable.

All patients have a septate or double vagina, and the two vaginas are often tilted away from each other with a wide intervaginal area.[8] As in 8 of our patients, the intervaginal septum may also be thick and fleshy. Division of the septum may then leave a gap between mucosa and mucosa and necessitate an inter-vaginal anastomosis to bring the mucosa together.[31,32] This surgery is best-performed in older girls, in whom the anatomy is more clearly delineated, thus reducing the risk of damaging the cervices while resecting the uppermost portion of the septum.[32,33]

Although not seen in our series, partially fused labia have been reported.[9,15] A short posterior midline cutback of the vestibular margin to expose both the urethral and the two vaginal orifices has also been reported.[9,11] In the light of our findings, this procedure would appear unnecessary, although its requirement during late adolescence or adulthood in some girls cannot be ruled out. A detailed prepubertal assessment and evaluation are mandatory in all patients.

Management of uterus didelphys

Uterine duplication with a double vagina appears to be invariable[8] although the exact terminology is also not clear. A uterus didelphys describes two uterine horns, each with a cervix, usually contiguous in the midline.[33] In girls with CPC, who have two hemi uteri and two cervices, with one on each side in the pelvis, it is not clear whether the comparison to uterus didelphys or the unicornuate uterus is most appropriate.[33] Although not performed in the present series, the exact anatomy could best be confirmed by coronal section MRI studies.

According to Grimbiz *et al*.,[34] unicornuate and didelphyic uterus have a similar effect on reproduction

and a poor pregnancy outcome (term delivery rates of around 45%). The pregnancy outcomes of untreated bicornuate and septate uterus are also poor (term delivery rates of only around 40%).[34] Between 21%[35] and 32.9%[34] of patients with uterus didelphys had a miscarriage. In addition, between 24%[35] and 28.9%[34] had a preterm delivery, with associated fetal growth retardation.

The possibility of uterine outflow obstruction due to cervical atresia or underdevelopment should also be kept in mind. In girls with ARM, Breech[33] emphasized the role of vaginoscopy before puberty. In CPC, the vaginas are usually not stenotic, and vaginoscopy is easy to perform. Vaginoscopy allows evaluation of the vaginal anatomy and documentation of the appearance, development, and position of the cervices and the presence or absence of mucus at the ectocervix (to infer cervical patency).[33] Antegrade or retrograde pertubation of the Müllerian structures during any open procedure can also confirm patency of the outflow tract.[33] In an earlier report,[36] we had described a girl with Type II CPC, not part of the present series, who had primary amenorrhea due to bilateral cervical atresia, necessitating bilateral hysterectomy due to advanced endometriosis of the uterus and fallopian tubes.[8] Gharpure[13] also performed bilateral hysterectomies in a girl with Type I CPC, although the indication for this procedure was not mentioned. Thus, there is a need for a comprehensive evaluation in the prepubertal period and a long-term management strategy.

In summary, in girls with Types I-III CPC, the colonic pouch ends in a fistula opening into the urethra or in the vestibule close to the urethral opening. As the fistula is closely apposed to the trigone and the urethra, it should be ligated low-down in the pelvis without attempting to dissect it completely from the lower urinary tract. The urethra opens either in a near normal position or at a "high" or hypospadiac level. UI, partial or complete, is frequent and appears to be due to a partially or completely open bladder neck and reduced bladder capacity along with a short, wide urethra. UI needs augmentation of outlet resistance and may require bladder augmentation for which a segment of the colonic pouch may be an alternative. All girls have a double uterus and vagina, the vaginas often being widely separated with a thick intervaginal septum. Division of the thick septum may necessitate an intervaginal anastomosis. The patients do not have "persistent cloaca" in its accepted sense and considering the wide vestibule, surgery for the UG component requires only division of the intervaginal septum, and if feasible, the septum between the fistula and vagina on each side. This leaves a wide vaginal introitus and a normal or hypospadiac urethral opening. A long-term management strategy is required throughout adult life for the associated gynecologic anomalies and their obstetric implications.

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