Pediatric Surgery International (2019) 35:263–269
https://doi.org/10.1007/s00383-018-4407-1

ORIGINAL ARTICLE

Early vaginal replacement in cloacal malformation

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Accepted: 18 October 2018 / Published online: 30 October 2018
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Abstract

Purpose We assessed the surgical outcome of cloacal malformation (CM) with emphasis on need and timing of vaginal replacement.

Methods An ambispective study of CM was carried out including prospective cases from April 2014 to December 2017 and retrospective cases that came for routine follow-up. Early vaginal replacement was defined as that done at time of bowel pull through. Surgical procedures and associated complications were noted. The current state of urinary continence, faecal continence and renal functions was assessed.

Results 18 patients with CM were studied with median age at presentation of 5 days (1 day–4 years). 18;3;2 babies underwent colostomy; vaginostomy; vesicostomy. All patients underwent posterior sagittal anorectovaginourethroplasty (PSARVUP)/ Pull through at a median age of 13 (4–46) months. Ten patients had long common channel length (> 3 cm). Six patients underwent early vaginal replacement at a median age of 14 (7–25) months with ileum; sigmoid colon; vaginal switch; hemirectum in 2;2;1;1. Three with long common channel who underwent only PSARVUP had inadequate introitus at puberty. Complications included anal mucosal prolapse, urethrovaginal fistula, perineal wound dehiscence, pyometrocolpos, bladder injury and pelvic abscess. Persistent vesicoureteric reflux remained in 8. 5;2 patients had urinary; faecal incontinence. 2 patients of uterus didelphys are having menorrhagia. One patient succumbed to sepsis at 7 months age. Renal functions remained normal in 16. One patient is undergoing dialysis.

Conclusion Early vaginal replacement in CM is feasible. Patients with inadequate introitus may suffer from menorrhagia. A regular follow-up is mandatory.

Keywords Vaginoplasty · Cloaca · Paediatric vaginal reconstruction

Introduction

The management of cloacal malformation (CM) is very complex. The initial management in the newborn period is usually a transverse colostomy. Further management depends upon the length of the common channel that requires a thorough assessment before contemplating pull through. While some advocate a total reconstruction in the same sitting especially in cases of low common channel less than 3 cm with vaginoplasty, urethroplasty and anoplasty, there are others who feel vaginoplasty can be done at a later date when hormonal influence allows for a larger vestibule and perineum with a more compliant patient. We thus aimed to assess our cases of CM both prospectively and retrospectively in the light of the need, timing and outcome of the vaginal replacement.

Methods

An ambispective study of CM patients was carried out including prospective cases from April 2014 to December 2017 and retrospective cases that came for routine follow-up during that period. Institutional Ethics clearance was not required as it was an observational study with routine care of the patients during admission for treatment and routine follow-up in the outpatient department. Early vaginal replacement was defined as that done at the time of bowel pull through. The timing of the various surgical procedures done and the associated complications were noted from the

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case records. The current state of urinary continence, faecal continence and renal functions was assessed.

Results

Eighteen patients with CM were studied with median age at presentation being 5 days (1 day–4 years). Table 1 depicts the associated anomalies. The pouch colon was complete in 1 and incomplete in 2. Three patients had associated Hydrometrocolpos, out of which one had pyometrocolpos. All babies underwent colostomy soon after birth. Three babies also underwent vaginostomy and two of these also underwent vesicostomy. Ten patients had common channel length of > 3 cm. All patients underwent posterior sagittal anorectovaginourethroplasty (PSARVUP)/ Pull through. Suprapubic cystostomy was done for one patient who underwent simultaneous extravesical ureteric reimplantation that was left for 3 months with intermittent clamping till good urinary flow was established post PSARVUP and vaginal replacement.

The median age at definitive surgery (PSARVUP/ Pull through) was 13 (4–46) months. The colostomy was closed 4–8 weeks later. The colostomy done was transverse in 14, sigmoid in 3 patients and pouchostomy with colorraphy in one patient of complete pouch colon following partial pouch colon excision that developed stomal prolapse. An emergency pull through was done to prevent further loss of colonic tissue. A delayed vaginal replacement was planned for her. The child recovered well, was discharged but in due course died of sepsis at the age of 7 months.

Out of the 10 patients with a long common channel, only PSARVUP was done in 3, the urogenital sinus (UGS) was left for later reconstruction in one case of pouch colon. This case died later due to sepsis. An early vaginal replacement was done in 6. In the remaining 3 in whom PSARVUP was done, the introitus was inadequate at puberty. One had a narrow persistent UGS that was repaired at puberty with a labial flap vaginoplasty to widen the introitus along with vaginal dilatation. The urethra is still opening on the anterior wall of the vagina and has been left as such as the child is continent for urine. Barrows flap vaginoplasty was done in one. One had a persistent vaginostomy till puberty through which occasional menstrual blood came out. The vaginostomy was surgically closed and vaginal dilatation was done under general anaesthesia as there was vaginal stenosis 2 cm from introitus. A vaginal polyp was identified on the posterior vaginal wall that later subsided on subsequent vaginoscopy following vaginal dilatation for 3 months. She is now on a regular vaginal dilatation program but has reached only 8 mm diameter size at age of 20 years. She has small follicular cysts in one ovary. She also has severe uncontrolled menorrhagia (13–15 days/month with intervening gap of 1–2 days after 6–7 days of bleed) for the past 1 year that has not responded to various hormonal therapies. Thus further surgery for redo vaginal construction is still not planned as she may need an intervention for the uterus didelphys.

In one patient while doing a PSARVUP with abdominal laparotomy, the length of the ileum taken for vaginal replacement fell short and appeared ischemic. Hence, a tubularized Gortex patch was applied at lower end of the vagina to prevent fibrosis between the urethra and the rectum. The baby kept having urinary tract infections. At 3-month follow-up, the distal colostomy segment was used for sigmoid vaginoplasty and the proximal colostomy was pulled down as the neo anus. The baby is doing well with continence for urine and stool both. Reduction clitoroplasty was done in 1 patient. One patient had presented with ambiguous genitalia with male-like urethralization (Fig. 1).

Six patients required vaginal replacement; 2 with ileum; 2 with sigmoid colon; vaginal switch in 1 and hemirectum was used in another for making the vagina (Fig. 2). The rectum was split into two halves longitudinally using staplers, preserving the vascular supply to each segment carefully (Fig. 3). The median age at vaginal replacement was 14 (7–25) months. Five of these cases were done during the period 2014–2017.

Table 2 depicts the various complications that occurred in the 18 patients and the treatment done for them. Four patients developed anal mucosal prolapse that required trimming. Urethrovaginal fistula developed in 1 patient that closed spontaneously after regular vaginal and urethral calibration. Two patients, who have attained puberty and are menstruating, are having menorrhagia. Both have uterus didelphys. One also developed a tubercular tubo-ovarian mass for which a pig tail catheter was inserted (Fig. 4). She is currently on antitubercular treatment.

Table 1 Associated anomalies with cloacal malformation

Associated anomaly	Cases ($n = 18$)	%
Solitary kidneys	2	11
Renal dysplasia	1	5.5
Vesicoureteric reflux	9	50
Partial sacral agenesis	2	11
Spina bifida (lipomeningomyelocele-1)	3	16.5
Pouch colon	3	16.5
Clitoromegaly	1	4.5
Septate vagina	4	22
Bicornuate uterus	5	26.5
Hydrometrocolpos	3	16.5
Ambiguous genitalia (Male-like urethra)	1	4.5
Ovarian cysts	3	16.5

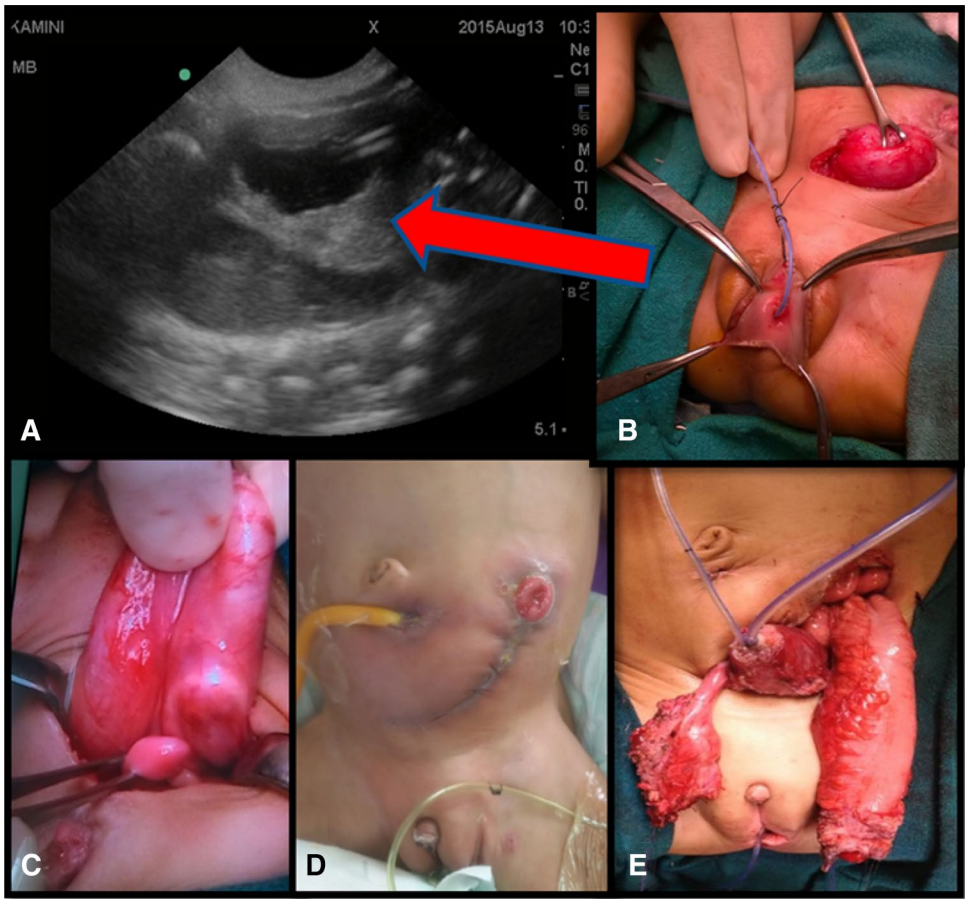

Fig. 1 An antenatal diagnosed left hydronephrosis patient had anorectal malformation and ambiguous genitalia at birth. Incomplete pouch colon was excised and end colostomy was made at birth. **a** Ultrasound pelvis demonstrating a cystic cavity with turbid material behind the bladder suggestive of hydrometrocolpos (arrow). **b** The male-like megalourethra was slit open into the perineum. **c** Exploratory laparotomy revealed a distended uterus, inflamed distended fallopian tubes, normal ovaries and vaginal agenesis. **d** Blocksom's vesicostomy done. **e** Left hemihysterectomy with left salpingectomy, left to right vaginal switch operation, left nephroureterectomy and end sigmoid colostomy pull through was done

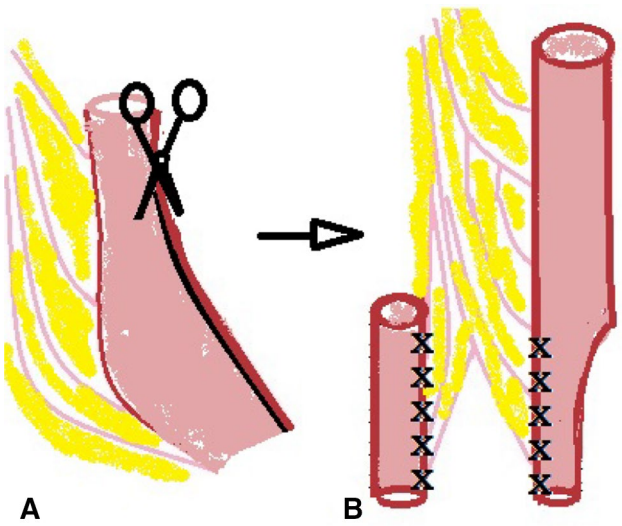

Fig. 2 Diagrammatic representation of splitting the rectum along the antimesenteric border. **a** Dilated rectum with good leach of blood vessels. **b** Mesenteric vessels split on either side to longitudinally split the rectum into two between the vessels and on the antimesenteric side to form two lumens for the neovagina and neorectum

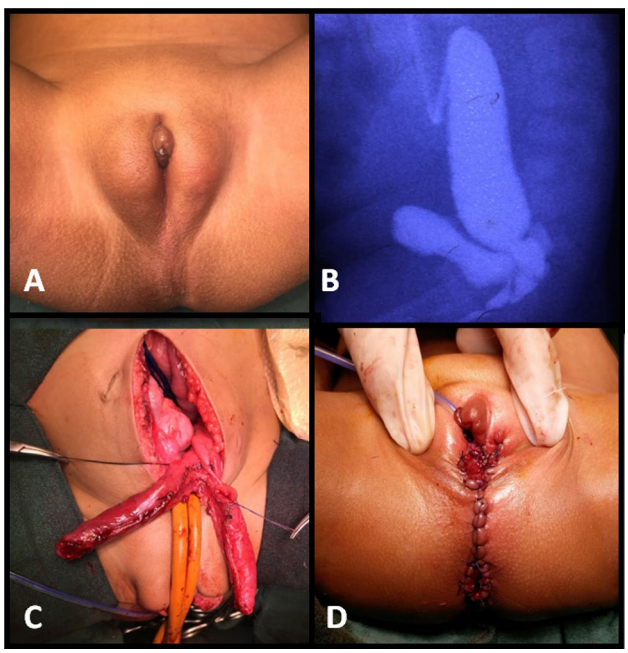

Fig. 3 **a** Clinical picture of a case of common cloaca subjected to early vaginal replacement. **b** Contrast study showing the common channel, bladder anteriorly, distal bowel superiorly opening between the two hemivaginae inferiorly. **c** Rectum split with staplers with neorectum on the right and neovagina on the left. **d** Completed repair

Table 2 Complications encountered with repair of cloacal malformations

Complication	Cases	Treatment
Per-operative and early post-operative complications		
Bladder injury	1	On table repair
Perineal wound dehiscence	3 (twice in one patient)	Secondary suturing
Perineal/vaginal infection	2	Perineal toilet
Pyometrocolpos	1	Vaginal douching with feeding tube
Urethrovaginal fistula	1	Vaginal and urethral calibration
Pelvic abscess	1	Laparotomy and drainage
Anastomotic leak following colostomy closure	1	Redo colostomy
Late complications		
Urinary infection	6	Antibiotics and Clean Intermittemt Catheterization (CIC). ureteric reimplantation, nephroureterectomy
Colostomy prolapse	1	Pull through
Anal mucosal prolapse	4	Mucosal trimming
Menorrhagia	2	Hormonal treatment
Vaginal stenosis	3	Vaginal dilatation-1 Barrows flap in 1 Labial flap vaginoplasty
Persistent urogenital sinus	1	Redo flap vaginoplasty (labial flap)
Anal stenosis	2	Anoplasty-1 Anal dilatation-1
Urethral stenosis	1	Urethral dilatation
Adhesive obstruction	2	Adhesinolysis-1 conservative-1
Tubo-ovarian mass	1	Pig tail catheter drainage
Mortality due to sepsis	1	–

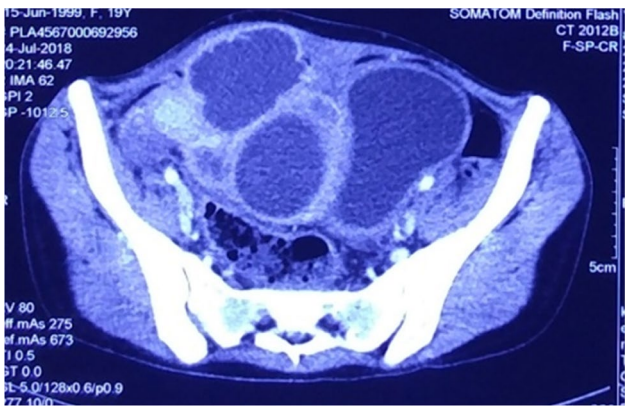

Fig. 4 CT scan depicting an inflammatory Tubo-ovarian mass situated between the bowel on the right and bladder which has been deviated on the left

The median follow-up period following vaginal replacement is 2 (1–12) years. The vaginal introitus is patent at last follow-up visits.

At a median follow-up of 8 (1–23) years, 5 patients had incontinence for urine and are on CIC. Three of these 5 patients had a common channel length more than 3 cm, 2 of these had associated spina bifida. Two patients had faecal incontinence and are on bowel management program. Both these had a common channel length more than 3 cm. Two patients had occasional fecal soiling with voluntary bowel movements.

Persistent vesicoureteric reflux (VUR) was found in 8, 5 received deflux; 1 underwent a ureteric reimplantation and 2 remained on chemoprophylaxis. One patient died due to sepsis at the age of 7 months. Renal functions remained normal in 16. One patient underwent left nephroureterectomy for a non-functioning kidney. One patient underwent extra vesical ureteric reimplantation. One patient is currently undergoing dialysis.

Discussion

The surgery for cloacal malformations is complex and requires expertise. Although the various options to repair the malformation have been described and the early outcomes following surgery are well reported, there is insufficient data on the complications encountered during follow-up and the long-term outcome. As the common cloaca patient grows older, the associated genitourinary anomalies require

attention. We aimed to study outcome of the surgical management of CM with an emphasis on the need and timing of vaginal replacement.

The initial diversion surgery in cloacal malformations is a transverse colostomy. However, in patients in whom sigmoid colostomy has been done elsewhere at presentation in the newborn stage, an early vaginal replacement may be considered utilizing the distal segment of the sigmoid colon for vaginoplasty at the same time as bowel pull through. Authors consider repairing the urethra, vagina and the anorectum at the same time beneficial in this complex malformation, without leaving the need for vaginoplasty in the future. This avoids the risk of fibrosis developing between the urethra and the rectum due to surgical intervention, which is likely to impede creation of space for vaginal replacement in future.

It has been wisely realized by surgeons dealing with common cloaca that the length of the common channel is an important determinant of the potential for urinary control, and predicts the extent of surgical repair. Patients with a common channel shorter than 3 cm can be repaired by PSARVURP route by most of the well-trained paediatric surgeons with a reasonably good prognosis [1]. Whereas, patients with a common channel longer than 3 cm (38%), usually require a laparotomy and have a much higher incidence of associated urologic problems [1, 2]. These complex technically demanding malformations should be repaired by surgeons with special training in urology. Also the functional results are not as good as those with a shorter channel. In Pena's series, 62% of the cases had a common channel less than 3 cm [1].

In cases where the common channel is more than 3 cm, a PSARVUP may be done in some cases. However, some of these girls may present with insufficient introitus at puberty to allow adequate menstrual flow. Three of our patients in whom PSARVUP had been done earlier had vaginal stenosis for which a labial skin flaps vaginoplasty, Barrows flap vaginoplasty and vaginal dilatation was done at puberty. Two patients still have persistent menorrhagia. Uterus didelphys may be a cause of increased menstrual flow in one of the girls who has a double cycle with apparently one hemiuterus bleeding followed sequentially by the other half. Hormonal therapy was also tried for a year for her but there was no relief. She is even ready for a hysterectomy to lead a better quality of life. The other girl is currently under antitubercular treatment for tubo-ovarian mass.

We have done an early vaginal replacement for six patients in our series. All these were planned at the time of the pull through. Though few complications were encountered but they could be easily identified and managed. Four out of these six patients are continent for bowel and bladder movements. When early vaginal replacement is planned, it is done usually with the time of bowel pull through which can be delayed till the child weighs around 8 kg and is nearing 1 year in age. Performing the urethroplasty after separating the vaginal tract meticulously without complete urogenital mobilization upto the bladder neck increases the chances of preserving urinary continence.

The technique used for vaginal replacement should have low morbidity, no mortality, prevent infections, prevent urinary incontinence, the opening should be adequate for efficient menstrual flow and should also serve as an appropriate conduit for coitus and married life without pain or discomfort. Three types of surgical procedures have been described for vaginal reconstruction for various indications in the paediatric age group, the inlay skin graft technique, the use of grafts (peritoneum, bladder mucosa, amnios) and the bowel vaginoplasty [3]. Amongst these, the sigmoid colon vaginoplasty is preferred. This is so especially in those cases of common cloaca that also requires a laparotomy for bowel reconstruction. The split-thickness skin graft is indicated only in cases of small reconstructions of distal vaginal tract as it requires lubrication, prolonged vaginal dilations or sexual intercourses to maintain patency [3]. In addition, it is prone to contracture, shortening, bleeding and dyspareunia. The other procedures such as amnion graft, pelvic peritoneum graft, Abbe-McIndoe procedure (perineal cleavage covered with a skin graft) or the Frank technique (mechanical dilatation) are best avoided in the paediatric age group as these tissues are very fragile and require mould insertions, vaginal stenting and dilatations [3].

Bowel vaginoplasty with an isolated ileum loop was first described in 1904 by Baldwin [4]. Subsequently, sigmoid conduit for vaginoplasty in the paediatric age was first described in 1911 by Wallace [3]. However, it was accompanied by a high mortality rate [3]. The procedure has been revisited over recent years and has now been used more often by the experts with good results [3, 5–11]. However, the colon may not be available in some cases of anorectal malformation with short colon, pouch colon or if the sigmoid colostomy has been fashioned. The second choice for bowel vaginoplasty is the ileum that is also preferred by some. The ileum may be used in cases where the pull through has already been done or in cases where the uterus is high up and there is not sufficient length of large bowel. This allows the sigmoid colon to remain as a reservoir for stool. In addition, in cases of complete pouch colon, the ileum is the only option available.

However, the ileum mucosa produces more abundant but less lubricating secretions than the sigmoid segment and is more fragile than colonic mucosa with higher chances of bleeding after intercourse [3]. The other advantages of sigmoid vaginoplasty is that the sigmoid colon is easy to mobilize, has a preserved blood supply, offers a more appropriately sized vaginal canal and permits to obtain an adequate length of 10–12 cm without problems with a low risk of

colitis and failure and offers possibility of having normal sexual function [1, 11]. Moreover, recently even laparoscopic vaginal reconstruction using sigmoid colon segment have been reported [12, 13].

We have used the vaginal switch procedure described by Pena in one case with good results. The prerequisite for a vaginal switch is that the horizontal width of both the hemivaginae should be more than the distance to bridge vertically [14, 15]. We used hemirectum in one case with satisfactory result. The rectum was dilated in this case and we split it longitudinally into two by splitting the leaves of the mesenteric blood supply into two (Fig. 2). This procedure has not been described earlier. Pena had described splitting of rectum keeping the distal blood supply for the neovagina and the proximal mesenteric blood supply for the neorectum that also depended on the intramural vasculature. However, this procedure depends on the presence of sufficient leach of blood vessels to divide into two.

The common cloaca patient does have mild clitoromegaly and a narrow vestibule. Two of our patients had abnormally large appearing clitoris and one had a male type urethra. One underwent clitoral reduction in infancy while in other; the megalourethra was split to bring the urethra to its normal location. The abnormal appearance of the genitalia also needs attention. If left untreated, this can lead to wrong gender assignment that may lead to psychological disturbance and even culminate in suicide later when delayed gender correction is done [16].

A careful follow-up following vaginal reconstruction with bowel has been suggested as there is a dismal risk of leiomyomas of the uterine remnants and adenocarcinoma arising in the intestinal neovagina reported in the literature [17, 18].

Many surgeons feel the ideal surgical time for vaginal replacement is after puberty [1, 19]. The reason being the patient is more motivated and compliant at puberty [1]. The onset of puberty has been reported to unmask several vaginal anomalies unrecognized in the prepubertal period such as vaginal atresia and cervicovaginal atresia [19]. In addition, hormonal impregnation at puberty ensures the development of an optimal vulvar tissue for anastomosis [1, 16]. This school of thought may be acceptable for cases with a non-functional or absent uterus. However, with anorectal malformation and cloaca, the uterus is functional and it is vital to provide an outlet before menarche and even earlier in presence of hydrometrocolpos. When the vaginostomy done for drainage of hydrometrocolpos is left too long, it hinders the growth of the vagina downwards with age as it does not allow the prepubertal or pubertal secretions to collect and there is a fixation effect of the vaginostomy pulling the vagina towards the abdomen.

We have reported our complications in Table 2 [20]. These complex procedures do have complications that need to be identified in time and dealt with. Similar complications like vaginal stricture or atresia, urethral strictures and urethrovaginal fistula have been reported earlier [1]. However, in Pena series these occurred before the introduction of the total urogenital mobilization (TUM). The TUM in early childhood provides good cosmesis and increases the urethral visibility on the surface for easy CIC if needed. It has been observed that the most girls with cloaca depend on CIC after TUM, especially when the common channel is long [21]. We have not used TUM in any of these patients. Although genitourinary surgical complications may be less after TUM, the incidence of urinary incontinence increases necessitating CIC. Bal et al. have also found that vaginal reconstruction avoiding dissection of the common wall between the vagina and bladder–urethra as well as avoiding TUM may help to preserve continence [16]. However, they preferred to perform the procedure at puberty. Nearly half of patients in Pena's series (54%) were continent of urine and 24% of these remained dry with CIC through their native urethra and 22% through a Mitrofanoff-type of conduit [1]. Upto three-fourth of the patients (78%) with long common channel (> 3 cm) required CIC compared to 28% with a short common channel (< 3 cm) [1].

Common causes of re- operations in common cloaca include colostomy prolapse, persistent urogenital sinus, atresia or stenosis of the vagina or urethra [1].

Only 11% of the patients in the series reported have faecal incontinence requiring bowel management. In Pena's series, 60% had voluntary bowel movements (28% of them never soiled, and 72% soiled occasionally) while 40% had faecal incontinence but remained clean when subjected to a bowel management program [1].

The limitations of the study include the relatively short follow-up of the cases that underwent early vaginal replacement. However, the longer follow-up of the patients that underwent PSARVUP compensates for this and emphasizes the need for early vaginal replacement in girls with a longer common channel to avoid complications during adolescence and young adulthood. Also the retrospective cohort of patients included only those that were in regular follow-up in the outpatient department. These thus included only those that were compliant to regular follow-up and those that had some problems especially after crossing the paediatric age group and puberty.

To conclude, early vaginal replacement in CM is feasible. Patients with CM with long common channel can be offered early vaginal replacement during the time of pull through with satisfactory outcome. However, these are complex procedures and should be done in a centre with a large experience of tackling these cases. PSARVUP is not sufficient for these girls as they tend to develop vaginal stenosis during puberty necessitating additional procedures. Uterus didelphys may be associated with menorrhagia that may be increased in cases with inadequate introitus subjected

to vaginal dilatation. A regular follow-up cannot be under estimated to identify the spurting urogenital problems and offer timely and effective treatment.

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