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Detubularized isolated ureterosigmoidostomy in a complicated common cloaca: A case report



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ABSTRACT

Urinary incontinence in a child secondary to a severe congenital anatomical disorder or due to complication of a previous surgery can be difficult to manage. Decisions can be especially hard when a redo procedure is being considered. We present one such case where a 6 year old girl previously operated for cloaca was brought with incontinence and after much consideration of options available, underwent a modified ureterosigmoidostomy to aid in her continence. The modification used was detubularized isolated ureterosigmoidostomy, described by Atta et al in 1996.

Key Words: Ureterosigmoidostomy, Atta pouch, detubularized isolated ureterosigmoidostomy, common cloaca.

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Introduction

Ureterosigmoidostomy (US) which was once used routinely by pediatric surgeons as a salvage procedure for failed bladder exstrophy patients, had largely been abandoned due to its high complication rate. Recently, it has regained popularity due to decreased incidence of complications with modifications of the procedure [1-6]. One such modification is the detubularized isolated ureterosigmoidostomy (DIUS), which was first described by an Egyptian surgeon Atta, hence also called the Atta pouch [6]. It works on the principles of detubularization. abolishing inhibitory reflex (RAIR), isolation of rectal and urinary segments and minimizing the contact of urine with colonic surface. We report the case of a 6 year old girl who was operated for common cloaca in another center, and presented with complications requiring a DIUS as a continent urinary diversion.

Case report

A 6 year old girl operated for cloaca as an infant presented with urinary incontinence and severe perineal excoriation. She was managed at another center in early infancy and had undergone a Posterior Sagittal Anorectal Vaginal Urethroplasty (PSARVUP) with a diversion stoma in a single sitting at the age of 2 months, followed by colostomy closure after a month. She now had urinary incontinence but was continent for stools. An otherwise intelligent girl, she was socially restricted and unhappy by the constant urinary leak and odor. The parents were in a state of desperation. On examination, her general condition was good she had thrived well. Abdominal examination was unremarkable except for the scar of colostomy closure. Perineal examination revealed a single large opening in the introitus about 1.5 x 1.5 cm in size, with skin tags and severe perineal excoriation (Fig. 1). Her neoanus was well sited with normal anal tone. Scar of PSARVUP was healthy.



Fig. 1. The single wide opening in perineum.

Blood investigations were within normal limits. Ultrasonography showed a left ectopic malrotated kidney. A genitogram showed two hemivaginae, and a small capacity bladder. On cystogenitoscopy, two orifices were seen deep in the introitus, no urethra, and small capacity bladder with virtually no bladder neck, both ureteric orifices were not seen, two hemivagina were seen with separate cervix in each. Since ureteric orifices were not visualized on cystoscopy, Computed Tomography Intravenous Urography (CT-IVU) Magnetic Resonance Imaging (MRI) of the pelvis was done in an attempt to confirm anatomical details. It showed a left ectopic malrotated kidney and communication of urinary bladder with common urogenital cavity. Right kidney was normal. MRI of the spine was normal. A diagnostic laparoscopy which showed a poorly developed bladder, bicornuate uterus with normal tubes and ovaries, bilateral ureters were seen coursing behind the cornu of the uterus on either side but not traceable beyond that. In spite of all investigations, we were unsure of the best option to offer the child, mainly to improve her quality of life. The only silver lining was that she had a continent rectum. The options considered were perineal reconstruction with urethroplasty and vaginoplasty followed by bladder augmentation with or without bladder neck closure. continent pouch, ureterosigmoidostomy or one of its modifications.

The choice of procedure took into account the available anatomy, prevalent social factors in her community, poor social acceptance of a girl with a stoma or a permanent urinary diversion, psychological benefit of dry perineum and the monetary benefits in terms of reduced diaper usage. We offered a DIUS or Atta pouch [6] for this child, after detailed counseling sessions with both parents and child. discussing all their options, consequences and preferences. An anorectal manometry was done which showed normal basal and squeeze pressures for age. In preparation for the child to be able to hold large quantities of liquid per rectally, we started training her by serial instillation of increasing amounts of saline into rectum. This lasted for almost 6 months, by the end of which she was able to hold a quantity of 350-400 ml of saline per rectally. This was done to ensure continence for liquids and improve capacity. She was also started on regular, vigorous perineal exercises.

Surgery was done by a Pfannenstiel incision. Left colon was dissected up to splenic flexure. An orientation suture was placed between left colon and lowest point on anterior surface of the rectum. This formed an inverted U-shaped rectosigmoid, approximately 30 cm in length (Fig. 2).

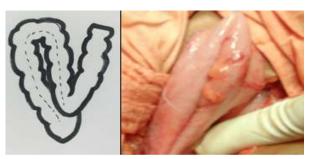


Fig. 2. Creating an inverted U-shaped rectosigmoid.

This loop of rectosigmoid was then completely detubularized along the anterior surface, as done during bladder augmentation. Posterior edges were anastomosed to form the posterior surface of the pouch. Both ureters were identified, dissected extravesically, hooked and divided, then anastomosed to the posterior surface of the pouch using a nippled technique (Fig. 3).

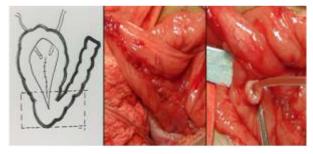


Fig. 3. An inverted U-shaped sigmoid pouch.

A raw surface was made in the posterior edge of the left colon opening and the left portion of the posterior wall of the rectal ampulla. The left colon was sutured to the rectal ampulla in line with the anorectal canal (Fig. 4).

Two 5 Fr infant feeding tubes were placed as ureteric stents and a rectal tube was placed in sigmoid colon beyond the pouch, all 3 brought

out per rectally and fixed. The anterior surface of the pouch was closed.

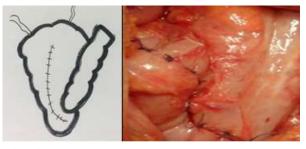


Fig. 4. The appearance of closed pouch.

Post operatively, the child was kept nil by mouth for 7 days, ureteric stents were removed by 10th day and the rectal tube was removed by 14th day. Initially she was only moderately continent for liquids but it improved well with time and with continuation of perineal physiotherapy. She was discharged on chemoprophylaxis and oral sodium bicarbonate.

On two-year follow up, she is "continent", has very occasional night time wetting and has had no urinary infections. More significantly, she is off diapers, dry, has no foul odor and no excoriation. She is able to differentiate between a urinary and a stool passage majority of the times. She has a significantly improved quality of life, goes to school regularly, mingles well with her friends and her parents are very satisfied.

We regularly follow her up with renal function tests, venous blood gas and yearly sigmoidoscopy. The future options we can offer this child are to either leave her as is, or do a perineal reconstruction under cover of the diversion.

Discussion

Ureterosigmoidostomy is the oldest form of continent urinary diversion first described by Simon in 1852. Though it remained hugely popular for almost a century, it started losing acceptance somewhere around the mid-20th century, due to the recognition of its late complications like electrolyte disturbances, ureterosigmoid anastomotic site stenosis, reflux. urinary tract infections with pyelonephritis and malignancy [1]. Recent evidence that most of these complications are due to unfavorable high-pressure conditions within the intact colon led to surgical modifications. The destruction of peristaltic integrity and refashioning of the bowel, along with anti-reflux procedures has led to the development of numerous innovative modifications [2]. There are more than 60 modifications of ureterosigmoidostomy at present [3]. The most commonly used modifications are the Mainz pouch II or the Sigma Rectum pouch [4] and the Modified rectal bladder described by Kock [5]. More recent modifications include DIUS by Atta et al. [6], ureterorectostomy using a modified Duhamel procedure by Fahmy et al. [7] and rectosigmoid bladder reservoir (RSBR) by Kanojia et al. [8]. Though long term outcome studies are available for most of the procedures, Kanojia et al. [8] are the only ones that have added a Health Related Quality Of Life (HRQOL) score making their outcome analysis a three dimensional one.

Atta et al from Egypt first described the modification of DIUS or Atta pouch in 1996, in bladder malignancy patients after radical In cystectomy [6]. the Atta pouch, detubularization of the whole rectosigmoid colon abolishes RAIR which is important for the patient to be able to hold large quantity of urine without feeling the urge to defecate, and also creates a low-pressure large-volume reservoir. Since the terminal colon is an intussusception into the pouch, it prevents reflux of urine from the pouch into the colon. This minimizes electrolyte imbalance and isolates urine and feces. There is also decreased electrolyte imbalance and carcinogenesis due to minimized exposure of colonic surface to urine.

Long term results of this procedure have showed continence during the day and night, with a satisfactory emptying habit [9], prevention of leakage even on full pouch and ability of patient to discriminate between urine and stool evacuations. However the rate of carcinogenesis can be commented upon only after further long term studies.

There can be complications like feco-urinary leakage, metabolic acidosis, febrile urinary tract infections and uretero-intestinal strictures, for which the child must be on regular follow up. The patient will also need to be on long term antibiotic chemoprophylaxis and oral sodium bicarbonate and will need regular monitoring with renal function tests, electrolytes, venous blood gas, ultrasound of the kidneys and yearly sigmoidoscopy.

This procedure has not been described for common cloaca so far in literature and the intent of this article was to discuss its feasibility and advantages in such cases with complications.

Conclusion

Detubularized isolated ureterosigmoidostomy or the Atta pouch is a rational modification of the conventional ureterosigmoidostomy that improves function after ureterosigmoidostomy by detubularization of the sigmoid and isolation of urinary pouch. The resultant separation of urine and feces proves very beneficial to the patient symptomatically and socially, while also reducing complications.

Compliance with ethical statements

Conflicts of Interest: None.

Financial disclosure: None.

Consent: Informed and written consent was taken from patient and her parents to publish this case report.

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