



ELSEVIER

INTERNATIONAL
JOURNAL OF SURGERYwww.theijs.com

Ureterorectostomy as a continent urinary diversion for complicated bladder exstrophy in children by using a modified Duhamel procedure: A case series

Mohamed A. Baky Fahmy^{a,*}, Abo Zid Aoud Mansour^b, Alaa Mazy^c

^a Department of Pediatric Surgery, Al Azher University, Cairo, Egypt

^b National Institute of Urology, Cairo, Egypt

^c Al Mansour University, Egypt

KEYWORDS

Duhamel pullthrough;
Bladder exstrophy;
Urinary diversion

Abstract *Introduction:* Whatever the method and timing of surgery, a high proportion of children with bladder exstrophy will continue to suffer from urinary incontinence. They face the options of urinary diversion to an external stoma or construction of a neobladder from bowel. This study describes a modified Duhamel's rectal pouch with a ureterorectostomy was carried out on 11 children who had a failed repair of bladder exstrophy.

Materials and methods: Ten boys and one girl, aged from 4 to 7 years (mean 5.5), had several unsuccessful operations for bladder exstrophy. All selected to have good renal function and no other anomalies, but were incontinent of urine and had a small contracted or prolapsed bladder. They underwent urinary diversion to the rectum using the Duhamel pullthrough technique, where the sigmoid colon was opened into the back of the anal canal above the dentate line, creating a rectal bladder and making use of the anal sphincter to control urine and stool. All were followed up for 24 months (18–27 months).

Results: In this selected group of patients there were no major operative or postoperative complications. Follow-up for 2 years revealed no deterioration in renal function, or electrolytes disturbance. They can hold up to 300 ml of urine and all patients are continent during the daytime with an emptying frequency of 3–5 times. Nocturnal wetting occur some 4–8 times per month with significant decrease with time. Two cases developed pyelonephritis but this was controlled with medical treatment.

Conclusion: Eleven children achieved effective urinary continence by ureteric diversion to the rectum using a modified Duhamel pullthrough technique. Two years follow up showed no complications, except bed wetting, but long term assessment is warranted.

© 2007 Surgical Associates Ltd. Published by Elsevier Ltd. All rights reserved.

Abbreviations: BE, bladder exstrophy; UD, urinary diversion; DP, Duhamel's procedure.

* Corresponding author.

E-mail address: mabfahmy@hotmail.com (M.A.B. Fahmy).

Introduction

Surgical reconstruction of the exstrophic bladder with local tissues is usually undertaken primarily or as a staged procedure. However nearly 10% of exstrophic bladders are of small capacity, and at least a further 30% of patients remain incontinent of urine even after a well performed staged reconstruction.¹ In Egypt the incidence of residual incontinence is high at about 40%, and most children remain diaper dependent, coming to consideration of internal or external diversion.² Ureterosigmoidostomy suffers from significant inherent complications including metabolic derangements, pyelonephritis, reduced growth, and particularly delayed carcinogenesis.³ Children with bladder exstrophy usually have a normal anal sphincter allowing construction of a neobladder from the rectum. 'Continence' and the lack of an abdominal stoma greatly contribute to a favourable body image. If done early the child could rapidly learn anal control for both systems, noticing little difference from the usual means of eliminating urine.

We report our initial experience with ureteric diversion to the rectum and bowel reconstruction with a modified Duhamel pullthrough technique.

Materials and methods

Ten otherwise normal boys and one girl with mean age of 5.5 years (4–7 years), had no anomalies other than bladder exstrophy and epispadias, were included in the study. Only patients with a normal upper urinary tract and normal calibre ureters were selected. The parents and eventually the patients were made fully aware of the long term specialized care that is required. Following 3–5 (median 3) previous surgeries as neonates and subsequently, 3 children had complete abdominal wall dehiscence and bladder prolapse, and 8 had a small contracted bladder without a bladder neck. Eight had an epispadias and all were incontinent of urine with total dependence on diapers. They had good renal function as assessed by blood urea nitrogen (BUN), creatinine clearance, renal ultrasound and renal nuclear scan, but one child had a grade II vesicoureteric reflux. Anal sphincter function was studied by contrast enema, anorectal manometry and anal muscle EMG using the anal plug and surface electrode, all cases proved to be normal with anal sphincter resting pressure of 50 mmHg (45–86), Maximum squeezing pressure 160 mmHg (150–390), which is normal compared with age matched controls.⁴ Five children also underwent voiding proctography, and only those who had normal findings were selected for diversion.

Duhamel pullthrough with ureterorectostomy

Bowel preparation was done 1 day preoperatively. Metrodazole and broad spectrum antibiotic were given during the operation. The child was placed in the Lloyd Davis position and the abdomen opened through a midline incision. After mobilization of the sigmoid colon the rectum was drawn up into the abdomen with stay sutures and a path developed behind it by blunt dissection down to the pelvic floor. The sigmoid colon was divided above the rectum.

The colon was passed behind the rectum, anastomosed to the anal canal and the posterior rectal wall above the dentate line and internal sphincter. An appropriate GIA autotapler was passed through the anal canal anatomising and dividing the colorectal septum and creating a common neorectum with a longer rectal spur than the one created for the Duhamel procedure used for treating cases of Hirschsprung's disease (Fig. 1). The two ureters were detached from the bladder and, using Goodwin's technique,⁵ were implanted in a subserosal tunnel on the lateral rectal wall below the upper edge of the rectum. Transanastomotic ureteric catheters were passed into the rectal space and were accessible through the anus. The proximal rectum was closed in two layers and abdomen closed leaving a pelvic drain. The residual urinary bladder was left in situ and excised subsequently at the time of genital reconstruction. Caudal block was done in 6 cases with 1 µg/kg of clonidine with ropivacaine for postoperative analgesia; with increasing the time to first analgesic request, it is effective in reducing pain scores, and increasing patient satisfaction.⁶ Oral feeding was established by the 4th day (3–6) and the ureteric splints were removed on the 10th day. The child was discharged home on the 13th day (12–15) postoperatively. Intensive toilet training to help the child control loose motions was started at the 3rd postoperative week, for a period of 1–2 months. Follow-up has been for 18 months for all and for 2 years for three children with assessment of serum electrolytes pH, blood urea nitrogen (BUN), creatinine and creatinine clearance. Data were entered and analyzed by SPSS 9.0.1 statistical package (SPSS, Chicago, IL, USA), and compared by paired *t* test. Data were considered significance if $p < 0.05$. Renal ultrasound was undertaken 3 monthly and a renal isotope scan 6 monthly. Rectal pouch capacity was assessed 3 months

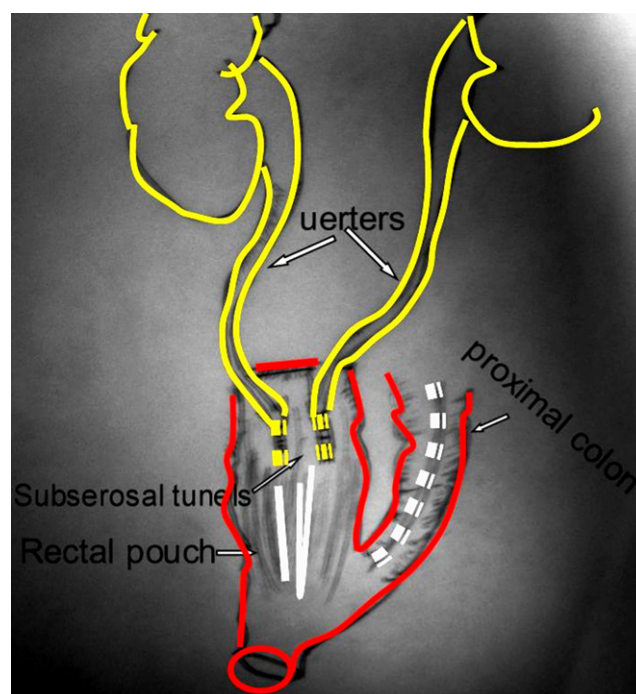


Figure 1 Duhamel Pull through with ureterorectostomy.

postoperatively by proctography with a soluble contrast medium. At each visit particular attention was given to the frequency of urinary evacuation and nocturnal bedwetting. The families were informed that the children would need proctoscopy or colonoscopy at least every 2 year or sooner if the motion contained blood.

Results

The mean operative time was 4 h (3–4 h). Blood loss was minimal, haemoglobin level and haematocrit value were maintained within the normal range postoperatively; blood transfusion was not necessary for any case. Postoperative recovery was uneventful and the children were allowed home 13 days (12–15) after surgery. Three monthly renal ultrasound demonstrated normal appearance. Blood gases, serum electrolytes, BUN and creatinine clearance were estimated twice during the stay in the hospital and at three monthly follow-up with maintenance of values within the normal ranges, but with significant improvement with time (Table 1). Soluble contrast proctography revealed an average rectal pouch capacity of 300 ml (from 280 to 360 ml) at 3 months, no reflux of the contrast to ureters was detected in any case (Fig. 2).

By the end of the 3rd month the children were able to retain motions for 3 h, which increased to 4 h by the 18th month. Initially bedwetting occurred some 7 times/month (from 6 to 8), but decreased to 4 times/month by the 18th month.^{4–6} Postoperative data are presented in Table 2. Three children had perianal skin excoriation and dermatitis, which was managed with local emollients. Two children had diarrhoea for 10–15 days postoperatively which was self-limiting. Another child required a constipating agent for 2 weeks to help with control of rectal content. Two children had a urinary tract infection with pyelonephritis 3 weeks after surgery and one of them required hospitalization and antibiotic management without recurrence in the follow up.

Discussion

Bladder exstrophy has an incidence of 1:30,000–50,000 live births.⁷ Reconstruction of exstrophy–epispadias complex remains one of the greatest challenges facing the paediatric urologist and the results are as yet uncertain. The goals



Figure 2 Postoperative proctography, showing the capacity of the rectal pouch.

of therapy include urinary continence with preservation of renal function, and reconstruction of functional cosmetically acceptable genitalia.⁸

Surgical reconstruction using local tissues is either undertaken primarily or as a staged procedure. The planned one-stage technique of Grady and Mitchell is promising with satisfactory short-term results, and staged procedures quote up to 88% urinary continence rates.⁹ However, nearly 10% of exstrophy bladders are unsuitable because of very small capacity, and another 30% of patients remain unacceptably incontinent even after a well performed staged reconstruction.¹ Others have low continence rates even following a staged approach, such that there is an appreciable need for a modality of internal diversion.⁹ In developing countries where early surgical reconstruction is not done routinely and there are limited technical resources, there is a high incidence of failure of bladder reconstruction for children with bladder exstrophy. Many, either primarily or

Table 1 Biochemical data of 11 patients followed for 18 months represented by median (minimum–maximum)

Biochemical data	Normal values	3rd month (n = 11)	6th month (n = 11)	12th month (n = 11)	18th month (n = 11)
pH	7.38–7.48	7.37 (7.35–7.4)	7.40 (7.36–7.41)	7.41 (7.39–7.42)*	7.41 (7.4–7.42)*
Bicarbonate (mmol/l)	22–29	23.5 (23–25)	24.5 (23–26)*	24 (22–26)	24.6 (24.5–23)*
Sodium (mmol/l)	135–150	139 (138–142)	141 (140–142)*	141 (140–143)*	141 (141–140)*
Chloride (mmol/l)	98–106	103 (102–105)	103 (102–105)	102 (100–104)*	101 (101–100)*
Potassium (mmol/l)	3.5–5	3.9 (3.7–4.1)	4 (4–4.2)*	4 (3.9–4.2)*	4 (4–4)*
Creatinine (μmol/l)	60–125	89 (80–95)	80 (72–85)*	76 (70–82)*	72 (69.5–68)*
Creatinine clearance (ml/min)	97–137	101 (95–113)	103 (97–112)	112 (102–119)*	116 (118–110)*
BUN (mmol/l)	1.8–7.1	4.8 (2.2–7)	4.1 (1.9–5.9)*	2.6 (2–3.1)*	2.2 (2–2.3)*

*Significant difference in relation to the results of the 3rd month, where $p < 0.05$.

Table 2 Rectal evacuation and bed witting of all cases, data as median (minimum–maximum)

	3rd month (n = 11)	6th month (n = 11)	12th month (n = 11)	18th month (n = 11)
Frequency to evacuate the rectum (h)	3 (2.5–4)	3 (3–4)	4 (3–4.3)*	4 (4–4.3)*
Bed witting at night per month	7 (6–8)	7 (6–8)	6 (5–7)*	4 (4–6)*

*Significant difference in relation to the results of the 3rd month, where $p < 0.05$.

following failed surgery, will come to consideration of diversion. Also the social taboos and culture in our locality often do not favour an external stoma and add to the need for an internal continent urinary diversion.¹⁰

Pahernik et al.¹¹ report their long-term results of conversion from a colonic conduit into a continent anal urinary diversion, as after conduit urinary diversion in childhood, some patients wish to have a later conversion to a continent diversion to avoid external appliances and to improve their quality of life.

Specific advantages such as an improved body image and a more independent lifestyle without external appliances seem to justify these more complex procedures.¹¹

The past 15 years have seen an increasing interest in continent diversion of the upper urinary tract. The concept of refashioning the bowel to serve as a urinary reservoir rather than a conduit is based on the original pioneering observations by Goodwin and others in the development of the augmentation cystoplasty.⁵ The destruction of peristaltic integrity and refashioning of the bowel has led to the development of many innovative urinary reservoirs constructed from bowel, using antireflux procedures to avoid upper tract sepsis and additional surgical techniques to achieve urinary continence.¹² Many of the 40 or so variants of continent urinary diversion presently used represent modifications of the parent procedures. The multiplicity of procedures reflects the fact that most remain less than satisfactory and the “best” continent diversion has yet to be devised. It is however becoming apparent that certain procedures are associated with lower early and late complications. Controversy surrounds which bowel segment is most appropriate for fashioning into a urinary reservoir, the best techniques for achieving urinary continence, and the best technique for prevention of reflux to the upper urinary tract.¹³ Ureterosigmoidostomy represents one of the oldest methods for achieving ‘continence’ for bladder exstrophy patients.¹⁴ However, the procedure carries the significant inherent complications of metabolic derangements, pyelonephritis, growth limitation and delayed carcinogenesis. There is an unacceptably high incidence of adenocarcinoma, reported at more than 10% in one series, developing adjacent to the ureterointestinal anastomosis in patients with urinary diversions that mix the urinary and faecal streams. Thus patients younger than 25 years of age with ureterosigmoidostomy have a 7000-fold greater risk of adenocarcinoma of the colon than the general population.³ Various innovative surgical techniques have been advocated for separating the faecal and urinary streams while still employing ureterosigmoidostomy principles. These operations can generally be considered together as rectal bladder urinary diversions. In each of these

operations, the ureters are transplanted to the rectal stump, and the proximal sigmoid colon is managed by terminal colostomy, or more commonly by bringing the sigmoid colon to the anal sphincter, using the sphincter for both bowel and urinary control. The principal concern is the calamitous complication of postoperative urinary and faecal incontinence, presumably occurring as a consequence of damage to the anal sphincter during the operative dissection.¹³

We have used the principles of the Duhamel pullthrough technique^{15,16} to internally divert urine for children who have failed to establish acceptable functional bladders after several operations for bladder exstrophy. This procedure is associated with minimal pelvic dissection and achieves a reasonable rectal reservoir. There is the theoretical advantage of avoiding a mix of urine and faeces, where the upper part of the rectal pouch will serve as a reservoir for urine and the faecal stream will come from the proximal bowel to the lower part of the rectum with preservation of the anal sphincter (Fig. 1).

Experimental studies comparing the absorption potential of rectum and sigmoid colon for electrolytes are in favour of the rectal diversion over the ureterosigmoidostomy which could possibly avoid hyperchloremic acidosis.¹⁷ Also a large number of adult patients have been reported with colon cancers following ureterosigmoidostomy, created after total cystectomy. However, there have been few reports of cancer in rectal bladder created instead of ureterosigmoidostomy to reduce the risk of cancer development.¹⁸

Short term follow up of these 11 patients showed that there is no deterioration of the upper renal tract and there was no electrolyte or acid-base disturbance. The procedure was very well accepted by the children and their families. The continent rectal pouch created by using the principles of the Duhamel pullthrough for Hirschsprung’s disease, with a long rectal spur and subserosal reimplantation of both ureters is feasible, easy to perform and is successful in the immediate short term with 100% daytime continence and 92% nocturnal continence and a low incidence of complications.

A continent rectal reservoir is of paramount interest for most of the parents in our locality, and this could be quite different from other countries.

Clearly long term follow up of a larger group of patients is required to assess the effect of this operation on linear growth and bone density, and in particular to assess the possible deleterious effect of urine on the rectal mucosa. Adenocarcinoma remains a major concern in young adulthood and effective monitoring must be integral to the follow-up of these patients.

Conflict of interest

None.

Funding

None.

Ethical Approval

None.

Acknowledgements

I would like to thank Mr D. Young and Mr A. Bianchi for their kind efforts in reviewing this paper, and Dr Susanne Krege, University of Essen, Medical School, Germany for her effort and observation of all of this work.

References

1. Chan DY, Jeffs RD, Gearhart JP. Determinants of continence in the bladder exstrophy population: predictors of success. *Urology* 2001;**57**:774–7.
2. Hammouda HM, Kotb H. Complete primary repair of bladder exstrophy: initial experience with 33 cases. *J Urol* 2004;**172**:1441–4.
3. Koo HP, Avolio L, Duckett Jr JW. Long term results of uretero-sigmoidostomy in children with bladder exstrophy. *J Urol* 1996;**156**:2037–40.
4. Cali RL, Blatchford GJ, Perry RE, Pitsch RM, Thorson AG, Christensen MA. Normal variation in anorectal manometry. *Dis Colon Rectum* 1992;**35**(12):1161–4.
5. Goodwin WE, Harris AP, Kaufman JJ, Beal JM. Open transcolonic ureterointestinal anastomosis: new approach. *Surg Gynaecol Obstet* 1953;**97**:295–300.
6. De Negri P, Ivani G, Visconti C, De Vivo P, Lonnqvist PA. The dose-response relationship for clonidine added to a postoperative continuous epidural infusion of ropivacaine in children. *Anesth Analg* 2001;**93**:71–6.
7. Molenaar JC. Cloacal exstrophy. *Semin Pediatr Surg* 1996;**5**:133–5.
8. Benson MC, Olsson CA. In: Walsh PC, Retik AB, Vaughan ED, Wein AJ, editors. *Campbell's urology*. 7th ed. Philadelphia, PA: WB Saunders; 1998. p. 3190–227.
9. Shaw MB, Rink RC, Kaefer M, Cain MP, Casale AJ. Continence and classic bladder exstrophy treated with staged repair. *J Urol* 2004;**172**:1450–3.
10. Elcoat C. Coping with stoma care in the community. *Practitioner* 1989;**22**(1469):776–9.
11. Pahernik S, Wiesner C, Gillitzer R, Stein R, Thurof JW. Conversion from colonic conduit into recto-sigmoid pouch (Mainz pouch II). *BJU Int* 2006;**97**:157–60.
12. Bastian PJ, Albers P, Haferkamp A, Schumacher S, Muller SC. Modified ureterosigmoidostomy (Mainz Pouch II) in different age groups and with different techniques of ureteric implantation. *BJU Int* 2004;**94**(3):345–9.
13. Stein R, Fisch M, Black P, Hohenfellner R. Strategies for reconstruction after unsuccessful or unsatisfactory primary treatment of patients with bladder exstrophy or incontinent epispadias. *J Urol* 1999;**161**(6):1934–41.
14. Simon J. Ectopia vesical operation for directing the orifices of the ureters into the rectum; temporary success, subsequent death, autopsy. *Lancet* 1982;**2**:568–70.
15. Duhamel B. A new operation for the treatment of Hirschsprung's disease. *Arch Dis Child* 1960;**35**:38–9.
16. Duhamel B. Retrorectal and transanal pullthrough procedure for the treatment of Hirschsprung's disease. *Dis Colon Rectum* 1964;**7**:455–8.
17. El-Mekresh MM, Shehab el-Din AB, Fayed SM, Brevinge H, Kock NG, Ghoneim MA. Bladder substitutes controlled by the anal sphincter: a comparison of the different absorption potentials. *J Urol* 1991;**146**(4):970–2.
18. Kotanagi H, Ito M, Koyama K, Sato K, Kato T. Colon cancer in rectal bladder. *J Gastroenterol* 2001;**36**(10):718–22.